THE PREVALENCE OF NEURAL TUBE DEFECTS AND THE OUTCOME OF MYELOMENINGOCELE IN CAPE TOWN

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1995

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A thesis submitted for the degree of

DOCTOR OF MEDICINE

UNIVERSITY OF CAPE TOWN

DECEMBER 1995

DECLARATION

I, Sandro Sabatino Buccimazza, hereby declare that this thesis is my own work and has not been presented for any degree at another university.

Signed by candidate

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Date: [illegible]

The work reported in this thesis was performed in the Department of Paediatrics and Child Health, University of Cape Town, Cape Town

To Fatima, Carlo and Paulo

ABSTRACT

This study was designed to document the prevalence of neural tube defects in Cape Town and to determine the outcome of children born with a myelomeningocele and operated on within the Neurosurgical service of the University of Cape Town.

The aim of the prevalence study was to document the frequency of neural tube defects (NTD) over a twenty year period in Cape Town (1973 - 1992) and to determine the effects of race, gender, maternal age, parity, and season of conception on the prevalence. Multiple sources of ascertainment were used, including all maternity hospital records, neurosurgical and spinal defects clinic data, as well as those from the Human Genetics Department and Fetal Abnormality Group.

The prevalence rates for NTD fluctuated between 1,74 and 0,63 per 1 000 births, but showed no significant trends over the twenty year period. Prevalence rates were highest for the white population group at 2,56 per 1 000 births compared to 0,95 per 1 000 for blacks and 1,05 per 1 000 for those of mixed ancestry. The higher rates in the whites, who are of British and European extraction and belong to the more affluent section of the community, would suggest that the possible effects of nutrition and infection are overshadowed by genetic factors. There was a female preponderance for both spina bifida (M:F ratio 0,89) and anencephaly (M:F ratio 0,67). The highest NTD rates were found at both ends of the maternal age range (<20 years and >35 years of age). The prevalence was highest at the extremes of birth order (1,65 and 1,58 for birth order 1 and >7, respectively, and 0,56 and 0,45 for birth order 5 and 6, respectively). A seasonal variation occurred which differed from that reported for the Northern Hemisphere and may reflect local climatic conditions.

The aim of the outcome study was to follow a group of infants who were operated on for myelomeningocele (01 January 1979 - 31 December 1985) and evaluate their outcome at five years of age, in an attempt to identify factors that may influence the quality of survival

and their outcome, and to utilise these findings to recommend altering and improving (where possible) the management of children born with myelomeningocele.

The paediatric neurosurgical service offered by the Department of Neurosurgery of the University of Cape Town is the only service of this nature for many hundreds of kilometres outside of Cape Town. More patients came from outside of Cape Town (65 vs. 53), were black (22 vs. 7), with parents of a lower level of education (43 vs. 18 with no secondary school education) and from social class III (40 vs. 18). The majority of mothers were in their twenties (68 vs. 50), their educational profile in keeping with their social class, and those receiving no formal education coming from outside of Cape Town (24 vs. 6).

Antenatal diagnosis of a myelomeningocele was only made in four patients who had surgical repair of this defect. The number of affected cases was higher in first borns (44/118) and the majority were delivered under medical supervision (101 vs. 17). Delivery by caesarean section was unrelated to head circumference (9/19 had a COH >90th centile), had a better chance of the child being continent of urine and a Modified Griffiths Quotient >70 but was unrelated to successful ambulation.

There was some correlation between the anatomical site of the myelomeningocele and the sensory level at birth (Correlation Coefficient 0,65), the majority being lumbar (65) or sacral (46) sensori-motor levels. Only one third (40 vs. 78) of the patients had their lesion closed within 24 hours of birth and half (59 vs. 59) by 48 hours, the majority coming from the Cape Town area (44 vs.15), the delays in referral of country patients being mainly transport related. Seventy seven percent of the infants had hydrocephalus requiring a drainage procedure all within the first year of life (91 vs. 27), 42,3% requiring only a single ventriculo-peritoneal shunt and 24,6% needed revisions of the procedure.

At five years of age three quarters of the children had a Modified Griffiths Quotient >70 (60 vs. 20), 15% (11) were in the borderline range (70 - 79) and of those with scores <70 four fifths (15) were mildly handicapped (50 - 69) and one fifth *(5)* moderately handicapped (30 -

49). The scores on the locomotor sub-test (mean 55) were lower than the other sub-tests, with personal/social (mean 82) and speech/hearing (mean 85) marginally higher than eye-hand (mean 80), performance (mean 79) and practical reasoning(mean 77).

The mean General Quotient for non-shunted patients was 80, those requiring one shunt were no different to those not shunted, but the group shunted more than once functioned significantly lower in all sub-tests and was related to central nervous system (CNS) infection.

Annual serial testing using a rapid developmental assessment test (DEI) showed a high correlation with the General Quotient obtained using the Griffiths Scales of Mental Development at 5 years of age and appeared to have predictive value for later intelligence (Correlation Coefficients ranging from 0,77 at 1year to 0,89 at 4 years).

The overall ambulation rate was 71,3% (57 vs. 23) and was related to the sensori-motor level of the lesion. The mid-lumbar (L3 and L4) lesions showed a delay in ambulation and an increased requirement for orthopaedic surgery (23 vs. 19). Children from the Cape Town area were ambulant earlier (24 vs. 13 ambulant by 2 years) which may reflect the availability of orthopaedic management.

Forty four percent were continent of urine by 5 years of age (35 vs. 45), the majority (25 out of 35) on the clean intermittent catheterization programme. The sensori-motor level (32 of the 35 were lumbo-sacral), level of intelligence (mean GQ 84 vs. 75), birth origin (17 vs. 8) and social class (19 social class I vs. 6 social class III) all played a role in urinary continence. Those living within the Cape Town area were more successful in becoming continent on the intermittent catheterization programme because of the proximity to medical and nursing expertise.

ACKNOWLEDGEMENTS

I wish to thank and acknowledge the help and support of

Professor C.D. Molteno, Professor of Mental Handicap, Department of Psychiatry and former Head of the Developmental Clinic, Department of Paediatrics and Child Health, University of Cape Town, who I have worked with since my paediatric registrarship in 1980. His enthusiasm, guiding hand and faith in my ability to complete this project has been my inspiration during many moments of dispair.

Associate Professor T.T. Dunne, Department of Statistical Sciences, University of Cape Town, for his endless patience and generous help in analysing my study data.

Dr M.M. Nelson, Department of Human Genetics, University of Cape Town, for allowing me access to departmental records and the records kept by the Fetal Abnormality Group, and for her inspiration and encouragement during the early phase of the study.

Professor J.C. de Villiers, former Head of the Department of Neurosurgery, University of Cape Town, for his support and encouragement during the study, helping to arrange financial support for the project, and allowing me access to departmental records, especially the earlier records which were unobtainable elsewhere.

Associate Professor H.A. van Coeverden de Groot, Department of Obstetrics and Gynaecology, University of Cape Town, for allowing access and granting me permission to use the Peninsula Maternal and Neonatal Service data bank.

The Medical Officer of Health of Cape Town for allowing me access to the population statistics.

The Department of Home Affairs for allowing me access to their records at their Regional Offices in Cape Town and Observatory.

My research assistant Mrs D.M. Phillips for her hours of searching for patients and meticulous data recording.

Mrs L.J. Daries, Maternity Records, Groote Schuur Hospital, for the many hours of retrieving folders and patient information.

Sister E. Burger, Spinal Defects Clinic, Red Cross War Memorial Children's Hospital, for patiently and efficiently co-ordinating all the follow-up appointments.

The Superintendents of Groote Schuur, Red Cross War Memorial Children's, Mowbray Maternity, New Somerset and Peninsula Maternity Hospitals for allowing me to include the study infants in this research project.

The South African Medical Reseach Council for their generous funding of the outcome study of this research project.

The medical staff of the Spinal Defects Clinic, Red Cross War Memorial Children's Hospital, for their willing co-operation.

Miss C. Bailey for her help with the tables and graphics of the results chapter of this thesis.

Last but not least, the children with myelomeningocele and their families for their unhesitating co-operation to be included in the research project and being so patient and tolerant with the testing. May they be the beneficiaries of my efforts.

PUBLICATIONS

The following publications related to this thesis

Buccimazza S.S., Molteno C.D., Dunne T.T, Viljoen D.L. Prevalence of neural tube defects in Cape Town, South Africa. Teratology 1994; 50:194-199.

Buccimazza S.S., Molteno C.D. Developmental evaluation of preschool children with myelomeningocele. Southern Afr. J. Child Adol. Psychiat. 1993; 5(1):25-29.

Viljoen D.L., Buccimazza S., Molteno C., Dunne T. The prevalence and prevention of neural tube defects in Cape Town. S. Afr. Med. J. 1995; 85(7):630-632.

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I INTRODUCTION

Myelomeningocele is considered to be one of the most complex birth defects compatible with meaningful existence (Liptak et al., 1985). The management of children with myelomeningocele has changed dramatically over the last thirty years. In the 1950's most affected children were left to die. The 1960's saw the advent of vigorous surgical treatment and by the 1970's, Lorber had introduced the concept of selection criteria for surgery (Lorber, 1971). Those children not operated upon were allowed to die. With early surgical treatment, Ames and Schut (1972), Shurtleff et al.(1974), and McLone (1981), have demonstrated that more than 80% of children survive. However, as a result of increasing survival, an awareness of the multiple problems faced by these children has arisen and this has led to new approaches to minimise their handicap. These problems include hydrocephalus and its management, motor and intellectual impairments, a neurogenic bladder with possible upper urinary tract damage, urinary and faecal incontinence, and psychosocial problems.

Approximately 80% of children with myelomeningocele will develop hydrocephalus, but shunt surgery allows most of the affected children to grow up with a normal or near normal intellect (Stein et al., 1975). Close attention to shunt function and the prevention and vigorous management of associated central nervous system infection can avoid significant intellectual impairment.

The level of limb paralysis is relatively fixed at birth, but virtually all children are able to stand alone or with the help of appliances (braces or platforms) by 2 years of age (Leonard and Freeman, 1981). Most children, even those with high lesions can be mobilised, either by being taught to 'walk' using appliances (crutches and braces) (Kupka et al., 1978) or with the use of a vertical wheeler (Gajjar and Price, 1988). Some children, particularly those with high lesions, require careful attention because of the development of a kyphosis and scoliosis (Samuelsson and Eklof, 1988).

During the past two decades many changes have taken place in the management of urinary incontinence. The most common urinary diversion is a tubeless cutaneous vesicostomy (Fernandes et al., 1994). Clean intermittent urethral catheterization (Lapides et al., 1972), the use of medication to alter bladder and sphincter tone (Fernandes et al., 1991), and the placement of an artificial urinary sphincter (Aaronson, 1986), have all been significant advances in providing continence. Today few children should reach adolescence still having significant problems with continence, and almost none should have significant renal impairment.

The odour of faeces because of soiling as a result of faecal incontinence causes one of the most serious psychosocial problems for a child with myelomeningocele. Today, the appropriate use of bowel stimulation and supportive dietary management and daily or frequent rectal washouts, allows the children to be socially acceptable with little or no faecal incontinence.

Secondary to the problems outlined above, psychosocial problems may develop and add to the handicaps faced by the child with myelomeningocele. Overprotection by parents often compounds the problem. Schooling, in particular mainstreaming, and the awareness of disability by the community, play a role in minimising handicap and should allow children to be productive adults.

The birth prevalence (Borman and Cryer, 1990) of neural tube defects (NTD) within countries and between countries shows wide variation. Not only is there a geographical variation (Leck, 1974), but secular trends cause cyclical peaks and troughs which are not necessarily paralleled between countries (MacMahon and Yen, 1971; Windham and Edmonds, 1982). Recently a general decline in the prevalence of neural tube defects has been documented in first world countries (Lorber and Ward, 1985).

Accurate statistics in South Africa prior to 1966 regarding their prevalence are not available. Earlier studies were of vertebral columns from the local black population in museum collections both in Johannesburg (Shore, 1930) and in East Africa (Allbrook, 1955), and these documented the prevalence of spina bifida occulta only. Data for Cape Town have been published in the past (Horner and Lanzkowsky, 1966; Singer et al., 1978; Cornell et al., 1983), the last being over a decade ago and limited. Horner and Lanzkowsky (1966) published a retrospective hospital-based study involving two maternity hospitals in Cape Town, while a five year survey (1975-80) by Cornell et al. (1983) was restricted by only involving patients served by the Department of Obstetrics and Gynaecology of the University of Cape Town. Singer et al. (1978) reported a three year period (1975-77) involving the whole population of the Cape Province. No long term studies are available regarding the prevalence of neural tube defects in Africa or South Africa and the previous local studies are limited.

Most studies on survival, outcome and follow-up of children with myelomeningocele emanate from industrialised countries (Lorber, 1971; McClone et al., 1981). Studies from non-industrialised or third world countries are sparse and concentrate on the neurosurgical aspects and survival (Brau et al., 1990) and on cultural aspects (Oyewole et al., 1985). Data from South Africa are limited and look at survival (Katzen, 1971 and 1981), artificial urinary sphincters (Aaronson, 1986) and orthopaedic management (Fraser et al., 1992) in myelomeningocele.

The lack of data regarding the prevalence of neural tube defects and the outcome of children born with myelomeningocele, not only locally and in South Africa but also in Africa, formed the rationale for this study.

AIMS

The aims of this study are:

- to document the frequency of NTD over a 20 year period in Cape Town and to determine the effects of ethnicity, gender, maternal age, parity and season of conception on the prevalence.

- to follow a group of infants who were operated on for myelomeningocele and evaluate their outcome at five years of age, in an attempt to identify factors that may influence the quality of survival and their outcome.

- to utilise the findings from the above to recommend altering and improving (where possible) the management of children born with myelomeningocele.

II HISTORICAL OVERVIEW

Neural tube defects are known to have existed since the dawn of history. Examples of spina bifida occulta have been reported in prehistoric human skeletons excavated in Morocco. On carbon-14 dating these date to 10 500 - 12 070 years ago (Epipalaeolithic period) (Ferembach, 1963). Similar findings in skeletons from the prehistoric, Roman, Anglo-Saxon, and Medieval periods in England have been reported by Brothwell and Powers (1968). Spinal defects have been described in the Japanese, the North and South American Indian, the African American and the American of European origin (Stewart, 1931), the Alaskan Eskimo (Stewart, 1953), the East African Black (Allbrook, 1955) and the South African Black (Shore, 1930).

Anencephalus, the most striking of these neural tube defects, in which there is virtual absence of the forebrain and the skull vault, was found in one of the earliest described Egyptian mummies. It was studied by Saint-Hilaire in 1826, and good illustrations by Meunier show without doubt that it is a human mummy, although it was found in the catacombs of Hermopolis where the sacred ape and ibis were normally buried (Brothwell and Powers, 1968).

One of the earliest detailed descriptions of spina bifida are attributed to Nicholas Tulp, who, in 1637 in Amsterdam, published his Medical Observations, an illustrated book recording observations. Amongst the 228 observations there is the first clear description of spina bifida. He described six cases of spina bifida, one of which was a child with an extensive lumbar myelomeningocele. He clearly recognised that the 'tumour' overlying the bifid spine contained nervous tissue and he strongly advised against surgical intervention. He also coined the name 'spina bifida', calling it 'spina dorsi bifida'. Tulp, who is depicted in Rembrandt's painting "The Anatomy Lesson", was a successful surgeon and held the post of anatomy demonstrator at the Surgeon's Guild in Amsterdam (Rickman, 1963 & Brocklehurst, 1971).

In 1714, von Ruysch distinguished between the paralytic and non-paralytic forms of spina bifida and suggested an association with hydrocephalus when he remarked that 'the two, allowing for the difference in site, were almost the same disorder' (von Ruysch cited in Doran & Guthkelch, 1961).

In 1761, Morgagni examined several specimens of spina bifida with hydrocephalus and anencephalus with spina bifida and recognised the relationship of spina bifida to hydrocephalus, by considering the excess of fluid to be the cause of the spina bifida 'tumour' (Bocklehurst, 1971). Cleland (1883) described in great detail dissections of a full-term infant with spina bifida infant with a lesion extending from the sixth thoracic vertebra to the first sacral vertebra and included an account of the deformity now known as the Arnold-Chiari malformation.

In 1877, Morton a surgeon in Glasgow, injected a solution of iodine in glycerine into the spina bifida sac and claimed success. Others followed, but efforts were soon abandoned (Doran and Guthkelch, 1961). Surgical excision was attempted in 1885 by Mayo-Robson who also tried to close the lesion, but this too gained little acceptance. A non-surgical traditional view on treatment was recommended by the London Committee of 1885 (Brocklehurst, 1971). After the turn of the century and the development of aseptic surgical technique, excision of the sac became accepted as the treatment method of choice (Doran and Guthkelch, 1961).

In 1886 von Recklinghausen published observations on the dissection of 32 cases of spina bifida, and concluded that spina bifida was the result of a disturbance in closure of the neural tube. Chiari in 1891 published his initial observations on herniation of the cerebellum as a result of hydrocephalus, and Arnold in 1894 described the pathological findings in an infant with a thoracolumbar spina bifida 'tumour', who died shortly after birth, where he observed the deformed brain-stem within the cervical spinal canal. It was in 1907 that Schwalbe and Gredig, pupils of Arnold, re-emphasised the close association

of the hind-brain malformations with spina bifida, and coined the term 'the Arnold and Chiari malformation' (Brocklehurst, 1971).

Weed (1922), injecting potassium ferricyanide solution into the ventricular system of pig embryos discovered the communication of the ventricular system and the subarachnoid space, and his further observations described cerebrospinal fluid production, the pathway of flow and the sites of absorption into the venous system. During this period Dandy had established that hydrocephalus could be caused by obstruction of the internal or external cerebrospinal fluid pathways and was not primary or idiopathic as previously believed (Dandy, 1919). Russell and Donald (1935), from their study of 10 cases of myelomeningocele, suggested that the site of obstruction in myelomeningocele was either the roof of the fourth ventricle, or around the brain-stem.

Various theories about the cause of the hydrocephalus in spina bifida have been put forward. Penfield and Coburn (1938) published their experience of the operative management of a single patient, who had had a thoraco-lumbar meningocele surgically treated some years previously, and subsequently presented with evidence of brain-stem compression. They concluded that the Chiari Type II malformation which they observed at a posterior fossa exploration was due to traction, as the cervical nerves ran in a cephalic direction and the thoracic nerves in a caudal direction, suggesting that the spinal cord was fixed during its development. This finding was supported by other studies at the time (Ingraham and Scott, 1943). Cameron (1957) postulated that the meningocele, or myelomeningocele, permitted a leak of the cerebrospinal fluid into the amniotic cavity affecting the pressure relationships in the fourth ventricle so that the roof foramina were not formed and the medulla and cerebellar vermis displaced into the cervical spinal canal by the abnormal flow of the cerebrospinal fluid. In 1958, Daniel and Strich showed that the great vein of Galen was elongated in the Arnold-Chiari malformation, which led Megison et al. (1967) to consider it as a possible cause for the hydrocephalus.

Ingraham and Scott (1943) operated on infants with myelomeningocele only if they had no serious neurological lesions and had survived for at least a year or 18 months from birth, and if their general condition was good. They had no difficulty in closing the spinal lesion, but there was little enthusiasm for such surgery because no adequate treatment for the associated hydrocephalus was available.

In 1951, methods of treating the hydrocephalus by procedures which bypassed the obstruction were developed. Matson (1951) described draining the CSF directly into the blood stream or some other body cavity. Within a few years, these procedures had improved by developing one-way flow valves (Pudenz et al, 1957; Spitz, 1959), and this revolution in the treatment of hydrocephalus formed the basis for the enthusiasm in the modern surgical treatment of the patient with myelomeningocele.

III LITERATURE REVIEW

III.1 PREVALENCE

III.1.1 PREVALENCE AND SECULAR TRENDS

III.1.1.1 GENERAL

A wide variation in birth prevalence of neural tube defects (NTDs) occurs within and between countries. Secular trends with cyclical peaks and troughs are also seen, but are not necessarily paralleled between countries.

Elwood (1973) reported an increase in the prevalence of anencephaly and spina bifida in Dublin from 1/1 000 live births in 1900 to 8/1 000 in the 1960's, with peaks in 1938 to 1941 and 1960 to 1961. He also compared this data with data from Boston in the U.S.A. The peak that occurred in Dublin in 1938-1941 resembled the peak in Boston from 1930-1934. A striking difference in trends in the prevalence of these defects is present over the years 1945-1965, the frequency declining in this area of the U.S.A. whilst it had increased in Dublin with the peak in 1960-1961. Suggestions with respect to aetiology are made. The possible role of the influenza pandemic of 1918-1919 and the changes in alcohol production and consumption during and after prohibition, are suggested as possible aetiological factors in the peaks in the 1930's.

Janerich (1973) reported on the prevalence of anencephaly and spina bifida in New York State from 1945-1971. He suggested that there was a regression of the peak seen in the 1930's and that it was continuing. The use of maternity hospital based data in these studies makes them difficult to interpret as major changes had taken place in the attitude and use by society of maternity hospitals during this period.

Windham and Edmonds (1982) showed a continuation of the decline in NTDs in the United States during the 1970's. They used data from three surveillance systems monitoring between 25 000 and 3,5 million births in the United States and showed clearly that the decline in NTDs was continuing, both for anencephaly and spina bifida. The overall NTD rate ranged from 0.49/1 000 births to 2,04/1 000 births. Snyder et al.(1991) analysed the deaths due to anencephaly published by the federal government in the United States from 1968 to 1987. It was purely a descriptive study looking at mortality rates and statistical analysis of the data was not carried out. They describe a declining trend for mortality rates as a result of anencephaly over this period for both white males (by 46%) and females (by 49,2%), which is far greater than that for non-white males (by 14,4%) and females (by 3,1%).

Trends during the mid 1970's in other cities in the United Kingdom, in particular Edinburgh and Glasgow, showed that the prevalence rate of anencephaly had been declining. Lorber and Ward (1985) reported from Sheffield in England that the annual number of NTDs born had fallen progressively during the 1970s and 1980s. They reported only one spina bifida birth in 1984 and a average of 18 were born per annum in 1968-1970. The rate of anencephaly had fallen from 2/1 000 births in 1968-1973 to 0,05/1 000 in 1981-4. This drop in the rate of NTDs was also seen in other health regions in England and Wales.

The EUROCAT Working group (1991) reviewed data from various centres in Europe and Britain from 1980-1986. This showed high rates for the British Isles (>3/1 000 births in Glasgow, Dublin, Northern Ireland and South Glamorgan) and lower rates for Europe (average 1,15/1 000 births). The downward secular trend was again demonstrated for the United Kingdom with the exception of Glasgow, South Glamorgan and Galway who all showed an upward trend in 1985-6. This may indicate that the bottom of the trough may have been reached and that the possible disappearance of NTDs is not a reality. In the European centres the NTD rate remained stable during the study period. A decline in the

rate of NTDs has also been shown in Hungary (Czeizel and Revesz, 1970) and in New Zealand (Borman and Cryer, 1993).

Secular trends for NTDs have been well demonstrated over a period of time in various geographical locations. Borman and Cryer (1990) have pointed out that comparing data from period to period and region to region is not recommended.

Peaks and troughs have been shown for the prevalence of NTDs in most first world countries (Elwood, 1973). Recently a decline in the prevalence of NTDs has also been documented in some of these countries (Czeizel and Revesz, 1970; Janerich, 1973; Windham and Edmonds, 1982; Lorber and Ward, 1985; Borman and Cryer, 1993). Reasons for the secular trends and the decline in the prevalence of NTDs have been suggested. It is however uncertain whether this decline is set to continue or whether the bottom of a trough may soon be reached (EUROCAT, 1991).

III.1.1.2 LOCAL

Neural Tube Defects (NTDs) are a major health problem in South Africa. Accurate statistics prior to 1966 regarding their prevalence are not available. Earlier studies were of vertebral columns from the local black population in selected collections in museums both in Johannesburg (Shore, 1930) and in East Africa (Allbrook, 1955), and reported on the prevalence of spina bifida occulta.

Shore (1930) reported a higher prevalence of spinal defects amongst blacks. He reported on abnormalities of the vertebral column in a series of 82 skeletons of Bantu natives of South Africa. He reported a prevalence of 3,6% for spina bifida occulta. Similar figures were later reported by Allbrook (1955) for the East African Negroes, who had studied 206

vertebral columns of East African ethnic origin. Levy and Freed (1973) confirmed these figures reviewing 5 363 radiographs of South African Negroes in Johannesburg.

Some studies have remarked that spina bifida and anencephaly are much less common among black babies than white babies. Simpkiss and Lowe (1961) reported on congenital anomalies in the African new-born from Kampala in Uganda and in 2 068 consecutive deliveries they reported no cases of spina bifida or anencephaly. Review of 5 498 hospital records of babies born from 1953 to 1955 yielded only two cases of spina bifida and two of anencephaly.

Khan (1965) reported from Nairobi that in 3 016 consecutive births, the rate of anencephaly was 0,99/1 000 births and the rate of spina bifida 1,66/1 000 births amongst East African Negroes. This figure correlates well with that of Singer et al. (1978) from Cape Town and Kromberg and Jenkins (1982) who reported on a large series (29 633 births) from Johannesburg. Similar figures were reported by Harrison et al. (1985) from Nigeria in a survey of 23 512 hospital deliveries in Zaria, showed a prevalence rate of 1,6/1 000 births for NTDs.

Stevenson et al. (1966) reported on congenital malformations, including neural tube defects, in a study of a series of consecutive births in 24 centres internationally, including Cape Town, Johannesburg and Pretoria. The South African data is flawed which questions the credibility of this data. In the study Somerset, Peninsula Maternity, Karl Bremer and Groote Schuur Hospitals from Cape Town, Queen Victoria Hospital from Johannesburg and Holy Cross Nursing Home from Pretoria co-operated. However, in the section regarding the List of Centres, Co-operating Hospitals and those responsible for the study, both Groote Schuur and Karl Bremer Hospitals are listed as Johannesburg hospitals, whereas they are/were both important maternity hospitals in Cape Town. The possibility that the data from these hospitals has been included in the data for Johannesburg exists. The figures given for Johannesburg throughout the text are higher than expected from the

one co-operating hospital (Queen Victoria Hospital) in the area and those for Cape Town lower, adding to that suspicion.

In the same year, Horner and Lanzkowsky (1966) undertook a pilot investigation to compare the prevalence of congenital abnormalities in the white and Cape Coloured (those of mixed ancestry) racial groups in Cape Town. They surveyed 6 502 infants delivered in two maternity hospitals under the aegis of the University of Cape Town. They reported a lower prevalence for anencephalus and spina bifida amongst whites than those of mixed ancestry, with a prevalence of 0,7/1 000 births for anencephalus and 0,4/1 000 births for spina bifida for whites and 0,8/1 000 births for anencephalus and 0,5/1 000 births for spina bifida for those of mixed ancestry. This was a retrospective survey and limited to two maternity hospitals and a small number of patients. The white population of Cape Town is largely of British or European extraction, where the prevalence of anencephalus and spina bifida was reportedly and still is higher than that reported by the authors. As most white patients would have delivered in private practice, unless the pregnancy was complicated or there were financial considerations, only a select group of white patients were surveyed. If anencephalus had been diagnosed in private practice it would not have been referred and if spina bifida was diagnosed in private practice it may have been referred directly to a neurosurgical unit and not a maternity hospital for further management. This may well account for the low prevalence reported for anencephalus and spina bifida amongst the whites in this survey.

Singer et al. (1978) reported on spina bifida and anencephaly in the Cape Province of the Republic of South Africa. They reported a prevalence of 1,59/1 000 births for both anencephaly and spina bifida for whites, a prevalence of 0,59/1 000 births for spina bifida and 0,12/1 000 births for anencephaly for those of mixed ancestry, and a prevalence of 0,69/1 000 births for spina bifida and 0,34/1 000 births for anencephaly for blacks. This figure for whites is similar to British figures at the time. The figure for those of mixed ancestry is higher than that previously reported and the figure for blacks is the first reported for the local black population. Many of the rural deliveries are home deliveries

and because of cultural beliefs the birth of a child with a congenital malformation may go unreported. Those delivered in rural private practice with anencephaly may not have been referred further.

Grace (1981) quoted rates of NTDs from Durban of 2,0/1 000 births for whites, 0,9/1 000 births for Indians, 0,67/1 000 for blacks and 1,81/1 000 births for those of mixed ancestry from his personal unpublished data.

Cornell et al. (1983) reported on the prevalence of neural tube defects in Cape Town for the period 1975-1980. The study was a retrospective survey of patient records in the population served by the Department of Obstetrics and Gynaecology of the University of Cape Town. They reported an overall rate for the five year period of 1,6/1 000 births for anencephaly, 2,0/1 000 births for spina bifida and total NTDs 3,6/1 000 births for whites, 0,33/1 000 births for anencephaly, 0,47/1 000 births for spina bifida and total NTDs 0,8/1 000 births for those of mixed ancestry, and a rate of 0,33/1 000 births for anencephaly, 0,22/1 000 births for spina bifida and total NTDs 0,55/1 000 births for blacks. Although the figures for the white population were similar to those in the United Kingdom at the time, the majority of the white population in this area would have delivered privately, as mentioned previously, and would not have been included in the search. The figure for anencephaly for those of mixed ancestry is higher than previously reported by Singer et al. (1978) and that for spina bifida lower. The figure for the black population for both anencephaly and spina bifida is lower than that reported by Singer et al. (1978). The black population in this area originates either from the Transkei or the Ciskei and there is often a to and fro migration of this population between these rural areas and Cape Town.

Ncayiyana (1986) reported on a retrospective review of 9 142 deliveries over a five year period from 1980-1984 in the rural Transkei district of Umzimkulu. The rate of anencephaly was 1,75/1 000 births, spina bifida 4,38/1 000 births and total NTDs 6,13/1 000 births. These figures are the highest yet reported for Blacks in this country.

The total number of deliveries in the area is unknown as a large percentage of the deliveries in the area were home deliveries (38%). Taking this into account and inflating the total number of deliveries the total NTD rate is closer to 3,79/1 000 births. This is similar to the rate described for Whites by Cornell et al. (1983) in Cape Town, but is still the highest rate described for blacks in this country. Long term monitoring of the prevalence of the condition in this area would be valuable as well as comparing this data with figures for the region as a whole.

Accurate prevalence rates for NTDs in Africa and South Africa are poorly documented. Studies that have been reported are over short periods of time (Horner and Lanzkowsky, 1966; Singer et al., 1978; Cornell et al., 1983; Ncayiyana, 1986).

III.1.2 SEASONAL PREVALENCE

In the quest of unravelling the aetiology of neural tube defects (NTDs), attempts have been made to relate the prevalence of these defects to the season of birth or conception. Initially this proved unconvincing but with time a trend has developed. Leck (1974) reviewed the earlier studies carried out in the United Kingdom and pointed out that the relationship of season to prevalence appeared to vary according to year, locality and type of defect.

McKeown and Record (1951) reviewed stillbirth data from the annual reports of the Registrar-General for Scotland, which had given the causes of stillbirths since 1939, and 930 consecutive births in Birmingham in 1940-7 with central nervous system malformations. They found that the prevalence of anencephalus to be significantly higher in children conceived in the period April to September and born in the period October to March, but no seasonal variation in the prevalence of spina bifida. Smithells and Chinn (1965) reviewing data from Liverpool showed an excess of winter births over summer

births for spina bifida but no seasonal variation in conceptions. Elwood and Nevin (1973) also showed a winter excess for both spina bifida and anencephalus births in Belfast, and showed a seasonal variation for spina bifida births but not so for anencephalus. On data from Europe, Czeizel and Revesz (1970) noted an excess of spina bifida (but not anencephaly) among spring conceptions during the 1960s. Tunte (1968) from Westphalia reported a peak in the prevalence of anencephaly among spring conceptions, but not so for spina bifida. Khoury et al. (1982) reviewed American data from the Birth Defects monitoring Program (1970-78) and the Metropolitan Atlanta Congenital Defects program (1968-79) and showed no seasonal changes for births of NTDs for singletons or multiple births.

Locally, Singer et al. (1978) reviewed the month of conception of 209 babies born with severe central nervous system defects in the Cape during 1969-1977. This showed a rise in conception in the second quarter of the year and a possible secondary peak in the last quarter. This data represents the whole of the Cape province, which is a vast area with a number of different geographical regions and population groups. Only numbers of cases are given and no rate has been calculated for conceptions or births of these defects. The possibility of this trend mimicking the monthly conception trend in the Cape is not excluded.

Ncayiyana (1986) in his study from Umzimkulu in the Transkei showed an increase in the number of NTDs delivered during the quarter October to December (Summer) out of proportion to the distribution during the other months of the year. He expressed these as a percentage of the total number of NTDs delivered in the period studied, but compared the data to all births per month for the same period also expressed as a percentage of the total.

A seasonal prevalence for NTDs has been suggested by some (Leck, 1974; McKeown and Record, 1951; Elwood and Nevin, 1973; Tunte, 1968; Czeizel and Revesz, 1970), but not

others (Khoury et al., 1982). Studies which show a seasonal prevalence show an increase in the prevalence of conception of a NTD during April to September (McKeown and Record, 1951). A seasonal prevalence for NTDs in South Africa has been suggested (Singer et al., 1978; Ncayiyana, 1986).

III.1.3 MATERNAL AGE

It has been suggested that the risk of having a child with a neural tube defect (NTD) is significantly greater for older mothers. Janerich (1972) showed by cohort analysis that the absolute risk of anencephaly was more directly related to the mother's year of birth than to that of the affected child. The suggestion made was that certain aetiological environmental factors affecting the mother early on in her life were affecting her offspring during her reproductive life. He suggested the 1918-19 influenza pandemic may have been a preconditioning event for the epidemic of NTDs which peaked in 1929-32. He also showed that the overall relationship of maternal age to the prevalence of anencephaly followed a U-shaped pattern. In a review of studies from the United States of America, Leck (1974) found a rise in rates near the end of reproductive life.

Smithells and Chinn (1965) in their study of 258 cases of spina bifida in Liverpool showed an excess of young mothers especially under 25 years of age. Bound et al. (1991) reviewed cases of NTDs in the Fylde in Lancashire during 1957-89. Data collected included 190 cases of anencephaly, 181 cases of spina bifida and 32 cases of cranium bifidum cysticum out of a total 116 282 live births and stillbirths during this period. The relative risk for anencephaly increased dramatically over 25 years of age and rose still further for mothers over 30 years of age. For spina bifida the maternal year of birth became a significant risk factor, and reaches a peak before 1950 and then declining. One of the suggestions put forward was that mercury, which was largely abandoned in the treatment of infants in the early 1950's, because of complications, may be an aetiological factor.

Local or regional data regarding the maternal age of infants with NTDs is unavailable. Data from the rest of Africa is scant. Of the 52 mothers described by Mabogunje (1990) from Nigeria, 27% were teenagers and 54% were in their third decade, but no further analysis of the maternal data is presented.

Studies suggest an increase in prevalence of NTDs at both ends of maternal reproductive life (Smithells and Chinn, 1965; Janerich, 1972; Leck, 1974; Bound et al., 1991).

III.1.4 SEX RATIO

Among individuals with NTDs a female predominance has been described in both the United Kingdom and the United States of America. Smithells and Chinn (1965) reviewing 258 cases of spina bifida from Liverpool found a female to male ratio of 1,2:1. Elwood and Nevin (1973) in their study from Belfast showed a female predominance, more so for anencephalus (26,3% male) than for spina bifida (41,1% male). James (1979) reviewing data from over 20 studies from different geographic areas in Scotland found a high female predominance. Windham and Edmonds (1982) in their extensive study in the United States, showed an overall female predominance although no sex ratio is quoted in their figures. This female predominance has also been supported by Janerich (1975). Khoury et al. (1982) also showed a female predominance in the United States, as well as a lower female predominance amongst nonwhites.

Myrianthopoulos and Melnick (1987) from Los Angeles supported the female predominance for NTDs in the United States, in the NIH Collaborative Perinatal Project, a prospective study of over 53 000 pregnant women and their offspring.

Dolk et al. (1991) as the EUROCAT working group reported a female predominance for NTDs, both in Britain and Europe, with male to female ratio of 0,65 and 0,89 respectively.

Seller (1987) has suggested that in populations where there is a high prevalence of NTDs, a female excess is more evident. Naggan (1971) from Israel, where the frequency of NTDs is low, has shown that more males are affected. It has also been suggested by Record and McKeown (1949) that females have more severe defects than males with NTDs and that an excess of females with NTDs are stillborn.

Local figures for sex ratios in NTD populations are scant. Levy and Freed (1973) in their study of spina bifida occulta in South African Blacks found that neither sex had a significantly higher prevalence of this anomaly. Kromberg and Jenkins (1982) in their study in Johannesburg found a male predominance in their 35 cases of NTDs (a F:M of 3:4 for their spina bifida group and 2,5:3 for their anencephalic group). Ncayiyana (1986), however, showed a female predominance for his 52 cases of NTDs in the Transkei (a F:M 3:1 for spina bifida and anencephaly). In both these studies numbers of cases are small, although the evidence from the Transkei, despite the small numbers, appears convincing. The local studies on NTDs of Horner and Lanzkowsky (1966), Singer et al. (1978) and Cornell et al. (1983) have not reported a sex ratio for the local NTD population.

Where sex ratios for NTDs have been documented, a female predominance has been reported - from the United Kingdom (Smithells and Chinn, 1965; Elwood and Nevin, 1973; James, 1979), from the United States of America (Windham and Edmonds, 1982; Janerich, 1975; Khoury et al., 1982; Myriathopoulos and Melnick, 1987), from Europe (Dolk et al., 1991) and in South Africa (Ncayiyana, 1986). Another South African study reported a male predominance (Kromberg and Jenkins, 1982). Local data from Cape Town has not been reported.

III.1.5 PARITY

Although a variation occurs with differences in maternal parity, it appears that this is geographic. Fedrick (1970) reported a higher frequency for anencephaly among the first born in young mothers in London, but in mothers over 35 years of age a parity of three or more was associated with an increased frequency. Leck (1974) reviewed data from the United States of America and reported the only common finding being that prevalence is low amongst second births. Naggan (1971) from Israel showed that there is an increased frequency of both anencephaly and spina bifida after the fifth child, and uniformly lower at lower birth orders. Hall et al. (1988) in a study from British Columbia showed no association with parity or birth order.

To date no reports regarding parity and the prevalence of NTDs have emanated from Africa or South Africa.

Data regarding prevalence of NTDs and parity or birth order is scanty but is suggestive of an increased prevalence amongst first borns (Fedrick, 1970) and again in the higher birth order (Fedrick, 1970; Naggan, 1971). Others have shown no increase in prevalence with birth order (Hall et al., 1988).

III.1.6 OTHER FACTORS

Socio-economic factors have been thought to play a role in the aetiology of neural tube defects, suggesting that they were more common in the offspring of families of low socio-economic status (Record and McKeown, 1949; Coffey and Jessop, 1957). Reviewing data from the British Perinatal Mortality Survey in 1958, Fedrick (1970) found the prevalence of anencephaly to be four times higher in social class V when compared to social class I and independent of regional variation, maternal age and parity.

Naggan and MacMahon (1967) showed a similar variation in Boston, where the risk for ward patients was twice as high as for private patients, and this was true for all ethnic groups except Jews. This trend has not been shown in either Israel (Naggan, 1971) or Hungary (Czeizel and Revesz, 1970).

Dietary factors have also been incriminated in the past. Following the association of the prevalence of neural tube defects and the range of geographical areas in the United Kingdom where the fungus Phytophora infestans (the cause of potato blight) occurs, it was suggested as an aetiological agent (Leck, 1974). Prolonged winter storage of potatoes, which promotes the growth of the fungus, and a high spring conception of neural tube defects correlates well, as does the fact that potato consumption is highest among the lower socio-economic groups. Although a steroid antibiotic, solanidine, which is produced in response to the fungus by the potato, has been shown to be lethal to chick (Renwick, 1972) and rat embryos (Kline et al., 1961), neither the fungus nor solanidine has been shown to cause congenital malformations.

The role of micronutrients in the aetiology of neural tube defects has existed for some time. The role of folic acid was originally put forward three decades ago (Hibbard and Smithells, 1965). Smithells et al.'s (1980) non-randomised trial of periconceptional supplementation with a vitamin and mineral preparation was difficult to interpret. The benefits of supplementation from possible risk of having a subsequent affected pregnancy was about one-seventh that in the unsupplemented women. A possible reason for the declining prevalence of NTDs in first world countries during the 1980's (Lorber and Ward, 1985; EUROCAT, 1991) could have been the widespread use of periconceptional vitamin supplementation following this study (Smithells et al., 1980), increasing awareness of the importance of diet both before and during pregnancy, and the wider availability of nutritious foodstuffs. However in some centres the declining trends had been noted earlier (Windham and Edmunds, 1982; Snyder et al., 1991) and in others the decline had occurred concurrently with massive local unemployment and a deteriorating social climate (Lorber and Ward, 1985).

In 1981 a second small randomised double blind trial of folic acid supplementation alone yielded inconclusive results (Laurence et al., 1981). The results of the multi-centred MRC vitamin study conducted in the United Kingdom have now shown that folic acid supplementation starting before pregnancy can reduce the risk of recurrence of neural tube defects (MRC, 1991). All these studies had concentrated on pregnant women with a prior NTD affected pregnancy.

Four observational studies from the United States have looked at pregnant women without a prior NTD-affected pregnancy. Three case control studies, one from Atlanta (Mulinare et al., 1988), one from Boston (Werler et al., 1993) and one from California (Mills et al., 1989) have shown different effects. Two studies have shown a 60% reduction risk in women who took a multivitamin supplement containing folic acid at least one month before conception. Mulinare et al. reported on 3 176 women of whom 14% used periconceptional multivitamin supplementation which continued through the first trimester, and Werler et al. on 3 051 women and the supplementation of folic acid ranged from 0,2-0,6 mg. and continued through to 28 days after the last menstrual cycle. However, Mills et al. showed that despite a multivitamin and folate supplement containing up to 0,8 mg. of folic acid and an adequate diet at least one month before conception through the first trimester, there was no protective effect, although numbers were small (579 women). In a prospective study in New England, Milunsky et al. (1989) has shown that supplementation with a multivitamin plus a folate supplement containing 0,1-1,0 mg. of folic acid and an adequate diet at least one month before conception through the first trimester had a protective effect (22 657 women).

The protective effect of folic acid has also been reported from other countries. Bower and Stanley (1989) from Australia has reported the protective effect of folic acid in a case control study from Western Australia. Although small numbers were involved (234 women), he compared the highest with lowest folate quartile and showed a 75% reduction in risk. From Hungary it has been reported that the results of a multivitamin/mineral supplementation (including 0,8 mg. of folic acid) among women who had not had a prior

NTD-affected pregnancy, showed a protective effect (Czeizel & Dudas, 1992). In a randomised controlled clinical trial involving 4 753 women planning a pregnancy, one group received a vitamin supplement (containing 0,8 mg of folic acid) and the other trace elements daily for at least a month before conception and until the date of the second missed menstrual period. Not only was there a reduction in the number of congenital defects in the vitamin supplemented group, but six cases of neural tube defects occurred in the trace element group and none in the vitamin supplemented group. Recently, it has been suggested that the metabolism of folic acid may differ in women at risk of having a neural tube defect affected infant (Wild et al., 1993).

For mothers planning a pregnancy to consume adequate amounts of folate-rich foods and to take folate supplementation while they are trying to conceive and for the first twelve weeks of pregnancy is possible. However, as most pregnancies are unplanned and most people are resistant to modify their eating habits a more general folic acid supplementation programme is necessary. It is now recommended that all women of childbearing age who are capable of becoming pregnant should receive folic acid supplementation for the purpose of reducing the risk of having a pregnancy affected with a neural tube defect (Oakley, 1993; Hitzeroth, 1993). The inauguration of such a policy may be difficult to achieve providing supplements to all these women and consideration to the fortification of staple foods, such as bread, flour and rice, with folic acid should be given (MRC, 1991; Sellers, 1994).

Low socio-economic class has been shown to be associated with an increased prevalence of NTDs by some (Record and Mckeown, 1949; Coffey and Jessop, 1957; Fedrick, 1970) but not by others (Naggan, 1971; Czeizel and Revesz, 1970). The role of the fungus Phytophora infestans has been incriminated as an aetiological agent in the past (Leck, 1974) but never proven. The protective effect of supplemental folic acid in preventing NTDs (Bower and Stanley, 1989; Czeizel and Dudas, 1992; Mulinare et al., 1988; Werler et al., 1993) and their recurrence (MRC, 1991) has been shown, but questioned by others (Mills et al., 1989). It has been suggested that folic acid metabolism

may differ in women at risk of having a NTD affected pregnancy (Wild et al., 1993). Following these studies it has been recommended that all women of child bearing age who are capable of becoming pregnant should receive folic acid supplementation (Oakley, 1993). Implementation of these recommendations will be difficult and the fortification of staple foods has been suggested (MRC, 1991; Sellers, 1994).

III.2 ANTENATAL DIAGNOSIS

The ability to diagnose a neural tube defect antenatally has been available for the past twenty years (Brock and Sutcliffe, 1972; Campbell et al,, 1972; Brock, 1976; Campbell, 1977). Initially, ultrasound scanning was used, and the first termination of pregnancy for a NTD following antenatal ultrasound diagnosis was performed for anencephaly in 1972 (Campbell et al., 1972).

Following the discovery that anencephaly and open spina bifida could be diagnosed using a raised maternal serum or amniotic fluid alpha-fetoprotein (Brock and Sutcliffe, 1972; Brock, 1976), population screening programmes were started in areas with a high incidence of NTD (Ferguson-Smith, 1983). Similar population screening programmes were commenced locally (Nelson and Coetzee, 1977; Grace et al., 1981). The test is best performed in the early second trimester and preferably before the twentieth week in pregnancy, before the normal rise in alpha-fetoprotein occurs, making diagnosis difficult. The antenatal diagnosis of an NTD, using this test, should be possible in at least 70% of cases. Some studies have reported better results (Campbell, 1977). Locally, most patients first attend antenatal care well into their second trimester of pregnancy, making it difficult to use maternal serum alpha-fetoprotein as a screening test (Van Coeverden De Groot, 1995). As with any screening test, there are other causes of a raised serum alpha-fetoprotein, and further investigations are indicated. Amniotic fluid is usually obtained to determine the level of amniotic alpha-fetoprotein as well as to check

for the presence of a nerve-specific enzyme, acetylcholinesterase (Report of the Collaborative Acetylcholinesterase Study, 1981). Using a combination of a raised alpha-fetoprotein and the presence of acetylcholinesterase in the amniotic fluid over 90% of the fetuses with a NTD can be detected. There is a risk of spontaneous abortion and fetal loss following an amniocentesis, but this is less than 1% (Scrimgeour, 1978; Simpson et al., 1979; NICHD National Registry for Amniocentesis Study Group, 1976).

Although the risk is small, an amniocentesis is an invasive alternative. In the past, using ultrasound, the diagnosis of a NTD has depended on the type of lesion. Anencephaly is diagnosable as early as 12 weeks' menstrual age, but usually confirmed between 14 and 20 weeks. Spina bifida presents a different problem. Scanning the spine is difficult and a diagnosis before 20 weeks rare (Campbell, 1977). Roberts et al.(1983), looking at 2 509 "at risk" pregnancies, reported a sensitivity of only 80% for sonographic diagnosis of spina bifida. Improvements in ultrasound technology and techniques, have resulted in an improvement in the sensitivity of the technique in the prenatal diagnosis of spina bifida (Richards et al., 1988; Hogge et al., 1989). Nicolaides et al.(1986) in a retrospective study, recognised that almost all cases of spina bifida were associated with aberrations of cranial anatomy, that could be diagnosed sonographically. The changes were secondary to the Arnold-Chiari malformation and include ventriculomegaly, disproportionately small head size, concave frontal bones (due to partial collapse of these bones) resulting in a lemon shape to the skull when viewed in the axial plane ("lemon sign"), and distorted anatomy within the posterior fossa of the skull, with anterolateral deviation of the cerebellar hemispheres around the cerebral peduncles giving a "banana-shaped" configuration. Some of these findings are dependent on the age of the fetus at sonography. The "lemon sign", for example, is present in nearly 99% of midtrimester fetuses with spina bifida but is less reliable after 23 weeks, when it is present in only 15% of cases. This was an important development, because when sonographic assessment of the spine and intracranial anatomy are considered collectively, the sensitivity of ultrasound for detecting spina bifida approaches 95%, similar to that of amniotic alpha-fetoprotein and acetylcholinesterase levels (Nicolaides et al., 1986; Campbell et al., 1987; Penso et al., 1987; Thiagarajah et al., 1990). A detailed fetal ultrasound is now the

preferred confirmatory investigation following the discovery of a raised early mid-trimester maternal serum alpha-fetoportein. In some centres, where mothers attend antenatal clinic services from early on in pregnancy, a detailed fetal ultrasound at 18 weeks, is done in preference to alpha-fetoprotein screening. Despite the improved technology greater operator experience is essential in maintaining the sensitivity of ultrasound for detecting spina bifida (Roberts et al., 1983). The training of ultrasonographers in detecting spina bifida is essential, but in areas of low prevalence it may be difficult to maintain their expertise. In Cape Town, no routine screening for NTD is carried out, unless indicated on clinical grounds or there is a family history of NTD or a history of a previously affected pregnancy. In these cases a detailed fetal ultrasound would be performed.

More recently, maternal serum alpha-fetoprotein levels have again come to the fore with the discovery that low levels in combination with unconjugated oestriol and human chorionic gonadotrophin levels (the triple test), can indicate an increased risk of the fetus having Down syndrome (Wald et al., 1992). Therefore, when high levels of serum alpha-fetoprotein are discovered using the triple test, further investigation is indicated to exclude the presence of a NTD.

The role of prenatal diagnosis and abortion of affected fetuses in developed countries when restricted to high risk cases had little effect on the overall births with neural tube defects, but as whole populations have been screened using not only maternal serum alpha-fetoprotein but high resolution ultrasound scanning the impact has been appreciable (Ferguson-Smith, 1983; Lorber and Ward, 1985). However, this alone cannot explain the declining trend in prevalence of NTDs in these countries (EUROCAT, 1991), as the decrease began in most places before prenatal diagnosis was available (Janerich, 1973; Windham and Edmunds, 1982).

Optimally the antenatal screening of pregnancies should be done before 20 weeks of pregnancy, allowing adequate time to legally terminate the pregnancy if required with a

minimum risk to the mother. The diagnosis of neural tube defects, in particular anencephaly and open myelomeningocele, can be made antenatally, using maternal serum alpha-fetoprotein (Brock and Sutcliffe, 1972; Brock, 1976) and/or acetylcholinesterase (Scrimgeour, 1978; Simpson et al.,1979). The use of the test after twenty weeks gestation diminishes as the natural surge in these products occurs. Antenatal sonography, initially not reliable in diagnosing myelomeningocele under twenty weeks gestation (Campbell, 1977) has been refined over the years and a detailed fetal sonograph is now used in preference to the alpha-fetoprotein screen at 18 weeks gestation in some centres (Nicolaides et al., 1986; Campbell et al., 1987; Penso et al., 1987; Thiagarajah et al., 1990). The success of the programme depends on the compliance of the attending public and the avilability of resources and expertise, the costs involved in running the programme and the equipment involved. Locally, most mothers attend antenatal clinic for the first time well into their second trimester making screening difficult (Van Coeverden De Groot, 1995). Although sonographic facilities are widely available in Cape Town and South Africa, high resolution fetal sonography is only available at a few large centres. Groote Schuur Hospital is the only hospital in the Peninsula Maternal and Neonatal Service to offer this service to the whole community under its care.

III.3 PERINATAL FACTORS

Until the development of prenatal diagnostic techniques, the presence of a myelomeningocele was only noted at birth. These techniques have allowed an assessment of the fetus to be made prior to delivery and have allowed some insight into separating the effects of the developmental lesion itself, of the intrauterine environment and the effects of labour and delivery on the lesion.

The role played by the intrauterine environment in the development of neurological damage in myelomeningocele has itself been questioned (Emery & Lendon, 1973; Osaka

et al., 1978). Using a pup model, Heffez et al. (1990) have suggested that amniotic fluid itself has a necrotic effect on the exposed cord and could act as an additional factor in the development of spinal cord injury.

The "stress of labour" is a well recognised entity and is due to altered placental perfusion during contractions and mechanical compression of the fetal head. Studies of vertex presentations show that the pressures applied to the cervix by the calvarium are 3-4 times the coincident intra-amniotic pressure. These forces increase after rupture of membranes and are repeatedly applied to the fetal head with each uterine contraction. The intra-amniotic pressure applied to the rest of the body, including the exposed cord, also varies with the magnitude of the uterine contraction. Biomechanical studies of breech presentations have not been performed, but traumatic lesions of muscle, skin and peripheral nerve in myelomeningocele patients following breech delivery have been described (Ralis, 1975). No mention was made of trauma to the neural placode. These injuries were attributed to local traumatic forces applied to the presenting breech.

Stark and Drummond (1970) reviewed their experience from the perspective of possible birth injury to the cord and brain. They found rupture of the myelomeningocele in 10% of cases after labour and vaginal delivery, and felt it likely that in "many others, the neurological deficit increased during delivery". Prompted by the variability in extent and composition of the neurological deficit seen in children with myelomeningoceles, they argued that the finding of a mixed upper and lower motor neurone lesion in 75% of patients suggested that trauma, as well as exposure to air and infection, might play a role in the eventual deficit.

Campbell (1977) used real time ultrasound studies on myelomeningocele fetuses in utero to determine whether ultrasound could reveal early evidence of paralysis. In two cases with major lesions the leg movement in utero was considered normal, but at birth, they were paraplegic, suggesting that labour and/or delivery might have resulted in additional injury. However, no intrapartum or postnatal data have been published for these fetuses.

Recommendations were put forward in 1984 for delivery management of a fetus with myelomeningocele by prelabour caesarean section. Nine cases were reported, four of which were diagnosed antenatally and delivered by lower uterine segment caesarean section (Chervenak et al., 1984). No antenatal ultrasonic data are presented to form an opinion of intrauterine limb movement as an assessment of neurological function antenatally, although neonatal limb function is presented in the four cases delivered by caesarean section.

Reports on the role of trauma during labour and delivery in the neurological function of patients with myelomeningoceles have been varied. Shurtleff et al. (1987) compared neurological function in infants delivered by prelabour caesarean section and postlabour vaginal delivery. High-level lesions incompatible with independent ambulation were seen in 15% of the prelabour section patients versus 39% of the postlabour-delivered children. Independent ambulators were seen after 48% of section deliveries and 14% after labour.

Other studies have shown no differences in outcome following either vaginal or caesarean section delivery. Bensen et al. (1988) were not able to demonstrate differences in developmental status or mortality in a group of 72 patients born either by caesarean section or vaginal delivery. Sakala and Andree (1990) retrospectively reviewed a variety of functions mediated by the spinal cord in vaginally and caesarean section delivered myelomeningocele patients and could not detect differences attributable to route of delivery. Hadi et al. (1987) reported that disruption of the arachnoid was not seen in vaginally delivered patients. In a study of 280 patients with myelomeningocele, Cochrane et al. (1991) have concluded that elective caesarean section did not offer a spinal cord or ambulatory advantage over vaginal delivery for those pregnancies presenting in a vertex fashion, but should be reserved for those presenting breech or who have other obstetrical indications for operative delivery.

The role played by the mode of delivery in the outcome of a myelomeningocele pregnancy is uncertain, partly because of the difficulty in knowing the functional neurological status of the infant prior to delivery. The role of caesarean section is as yet unsettled in myelomeningocele, except in a breech presentation and where other obstetric indications exist (Cochrane et al., 1991). Some studies have advocated the caesarean section route of delivery as being advantageous to the infant's neurological status (Campbell, 1977; Chervernak et al., 1984; Shurtleff et al., 1987), whilst others report it not advantageous (Bensen et al., 1988; Sakala and Andree, 1990; Cochrane et al., 1991).

III.4 POSTNATAL FACTORS

III.4.1 DEFECT CLOSURE

The arguments concerning whether or when to close the defect in a case of myelomeningocele, have vexed clinicians for many years. The case for closure in all myelomeningoceles has been put forward as have arguments for selection prior to closure.

Without surgery, the chances of survival of a child born with a myelomeningocele were slim. Laurence (1966) described the natural history of untreated spina bifida cystica in his study of 426 cases of spina bifida cystica and encephalocele from South Wales, born between 1956-1962, before active surgical management for the spinal lesion was readily available in the area. He followed the survivors for between two and a half and nine and a half years. He showed that in the absence of surgery only 16% of these cases survived, and that if one excluded the stillbirths and early neonatal deaths as potentially not salvageable, only 60% were salvageable with surgery. In a further follow-up of these patients, 15% of the liveborn myeloceles survived to 4 years of age, 70% were moderately or severely physically disabled and 34% had an intelligence quotient above or within the normal range. Only 4,25% of liveborn meningoceles and myeloceles survived with normal or near normal physical capacities and intelligence quotients above 85 (Laurence & Tew, 1966).

Since 1950, the role of surgery has played an ever increasing role in the newborn with a myelomeningocele. Chambers (1950) from Indiana reported on improved function and survival in ten babies born with myelomeningocele who were operated on soon after birth. A delay in surgery of 9 hours to 16 days post delivery was specified in only 7 cases, with no delay specified for the remainder. The 'good' results claimed were for the 7 cases where the delay was specified. These results although not conclusive suggested that early closure of the defect improved the outcome of the infant. Doran and Guthkelch (1961) in their retrospective study of 243 cases of myelomeningocele from Manchester,

136 of whom were closed between 3 and 12 months of age, concluded that the repair of the defect did reduce the risk of death from meningitis and rarely caused any deterioration in neurological status. They also felt that early operation (within 48 hours) was not advantageous to the neurological deficit.

In the next few years the focus shifted beyond the survival of the infants receiving early surgery, to other benefits arising from the closure of the defect. Guthkelch (1962) reported on 97 patients who were operated on at varying intervals post delivery (range 0 - 24 months), 7 within the first week of life and a further 13 within 1 month. He could find no difference in muscle power between children operated on early or late, except that his 'early' cases extended well beyond the 48 hours suggested by others (cf. Sharrard et al., 1963). The following year, Sharrard et al. published their controlled trial of immediate and delayed closure in spina bifida cystica. They randomly allocated 40 infants admitted within 48 hours of birth into two groups either for immediate closure of the defect or not. Despite the limited sample size their results suggest that early closure of the defect (within 48 hours of birth) reduces mortality, local sepsis, meningitis, ventriculitis, the duration of hospital stay, and the extension of muscle paralysis present at birth. Their recommendation for the latter is that a temporary neuropraxial type of lesion of spinal cord or nerve roots develops during labour, and that these consequences are reversed by the immediate closure (Sharrard et al., 1963; Zachary, 1965). Sharrard et al. (1967) published a larger series of 526 infants born between 1955 and 1962 with open myelomeningocele and followed-up in 1966. The spinal lesion was closed within the first four days in 274 infants and the others were treated by initial conservative management. This larger series showed that the mortality of operation during the first four days of life (48,8%) is significantly less than that of conservative management (67,8%). Infants who had closure of the spinal lesion on the first day of life had the best chance of survival. The improvement in limb function shown in their previous study was confirmed, especially if closure was performed on the first day of life, but operation within 48 hours of birth gave similar results. Brocklehurst et al. (1967) challenged this finding. They reported on the outcome of 25 consecutive infants with open myelomeningocele,

who were studied before and after closure. Clinical assessment postoperatively indicated that although there may be apparent improvement in voluntary function in some cases within the first few weeks, there was no significant increase in useful leg function compared with preoperative levels. At three months of age, 19 out of 22 survivors were unchanged. The number of infants is small and it is of note that 15 of the 25 infants had sensory levels above L1 preoperatively and would have in any case been paraplegic. Smyth et al. (1974), in their study on delayed closure substantiated this finding. They conducted a controlled trial of 99 patients with a lumbar or lumbosacral myelomeningocele, which was not leaking and was not complicated by hydrocephalus at birth (head circumference <38 cm.) from 1965-1970. The children were randomly allocated to one of two groups: early closure (<48 hrs.) or delayed closure (>48 hrs.). No difference in muscle power could be shown between the groups. Forty two (42,4%) died within the first year of life, especially the infants with reduced muscle power (the higher lesions). Boston and Wilkinson (1979) suggested that an initial non-surgical approach did not result in any deterioration. In a retrospective analysis they compared the outcome of 88 patients with thoracolumbar myelomeningoceles with a neurological deficit below L1, born in 1976-1977 and not operated on, with 76 patients with the same condition from 1964-1971 who were operated on. Mortality was found to be significantly lower in the surgically treated group only over the age of three months. Fewer of their infants in the untreated group developed progressive hydrocephalus and the neurological status of the survivors at a year of age was the same in both groups. Ventriculitis occurred in 26% of the surgically treated group and of the 50 with hydrocephalus, 16 developed ventriculitis, while none of the 11 without hydrocephalus developed this complication. They suggested that as early surgery increased the incidence of progressive hydrocephalus and ventriculitis, and might increase survival of more disabled infants, non-surgical treatment was justified since survivors are no worse off.

Laurence (1964) published his findings on 407 cases of spina bifida cystica born between 1947 and 1956 and treated in the London area. Three hundred and sixty eight had myeloceles, of whom 160 had surgery, at varying times some over three months post

delivery. All cases were followed up in 1958 and age at follow up varied. The expected survival to the age of 12 years was 29%. Of the 235 cases who developed hydrocephalus, 28% were present at birth and the majority had lumbar defects. Of the 185 cases that died, all but three had hydrocephalus and infection was the commonest immediate cause of death.

Beks et al. (1966), reviewed 162 children with spina bifida seen in their clinic from 1954 to 1965 (148 with myelomeningocele and 14 with meningocele). The majority had lumbosacral lesions. Surgery was carried out on 133 of the patients, of whom 122 survived, Seventy of the 122 developed hydrocephalus, of whom 56 were treated using a variety of operative procedures, following which 9 did not survive. At follow-up a further 15 had died. Of the surviving 98 patients data on 95 were available. Thirty six (34,8%) were not ambulant, 42,6% of the children who were old enough to be continent (75) had faecal and urinary incontinence, and only 37 were tested psychologically (20 who had hydrocephalus). They suggested that the intellectual development of children with spina bifida is practically normal, provided the hydrocephalus which may develop can be treated early and successfully, and that in view of their findings children suffering from spina bifida must be treated. Their findings were optimistic, but no differentiation was made on the outcome of the children with meningocele, and only a small sample of the children were formally tested psychologically.

Matson (1968) stressed that in non-epitheliazed forms of spina bifida cystica the decision to operate and the best time to operate were two different problems. He advised custodial care where there was total absence of nerve function below the upper lumbar levels. He also noted that in his opinion excision of the myelomeningocele never brought about a recovery of totally paralysed limbs or sphincters.

Rickham and Mawdsley (1966) from Liverpool showed that if the child with spina bifida cystica was actively managed surgically, the majority would survive infancy. They studied 157 liveborn children with spina bifida cystica born between 1960 and 1962,

100 were operated on, and of these 71 survived to three years of age. Of the 57 cases not operated on only 2 survived to the same age. Further follow-up of these infants and those born between 1963 and 1965 in Liverpool showed that 80% of those children who survived the first 24 hours of life came to surgery and that 70% of these children were surviving to school age and beyond (Mawdsley and Rickham, 1969).

Heimburger (1972) from Indiana, in a long-term follow-up study, found a high mortality rate, especially if surgery was delayed beyond 48 hours after birth. He reviewed 104 patients treated between 1950-1955 of whom 97 were followed-up during 1968-9, the survivors ranging in age from 13-19 years. Seventy one of the 97 had had a surgical repair of their myelomeningocele, 28 within 48 hours of age. Those who were over 48 hours old at referral had a surgical repair of their lesion within a day or two of admission. None of the infants' whose parents refused surgery survived, the majority dying within the first month. Thirty seven (38%) of those operated on survived to follow-up. The highest mortality and highest incidence of hydrocephalus was present in the group referred 1 week to 1 month post delivery. Twenty eight of the 37 survivors were attending school, 22 in age appropriate classes. Of the nine not attending school, 4 were mentally retarded. He concluded that early closure was optimal for survival and that closure should be delayed for referrals between 1 week and 1 month post delivery until after 1 month of age, by which time the majority would have died without surgical intervention. Because of the higher incidence of hydrocephalus in the referrals who were surgically corrected later (1 week to 1 month), he suggested that the development of hydrocephalus was influenced by surgery. However, he did not comment on whether the hydrocephalus was present prior to surgery, which may have influenced the delay in the original referral of the patient, as the incidence of hydrocephalus in both the operated group (70,4%) and the unoperated group (72,6%) was almost identical.

With the increase in survival following surgery in these patients, handicaps also increased. Laurence (1974) compared data from South Wales on 113 aggressively surgically treated cases born between 1964-66 (57 survivors) with the data from the unoperated cases in his

1956-1962 series (54 survivors). Survival improved from 17% without to 50% with aggressive surgery. However, the number of handicapped, both physical and mental, almost doubled.

Adams et al. (1985) from Atlanta looked at the survival of infants with myelomeningocele. They reviewed data on 154 infants born with the condition from 1972 to 1979. They found that there was an overall survival of 57% to one year, more surviving their first year in the late seventies than the early seventies, if their defects were closed and low on the spine. They did not include the treatment given to the infants nor any changes in treatment policy over the 8 year period. It is noteworthy that the number of infants decreased over the years, 75 (48,7%) were born between 1972 and 1974, 50 (32,5%) from 1975 to 1977 and 29 (18,8%) from 1978-1979, suggesting a declining secular trend. The timing also coincided with the use of alpha-fetoprotein in the antenatal diagnosis of neural tube defects, which was started in Atlanta in 1976, but the authors note only one case of open myelomeningocele was diagnosed antenatally by 1979 using the screen.

McLaughlin et al. (1985) from Seattle showed that there was a increase in the survival of patients having early surgery for a myelomeningocele. They reviewed 212 infants over an 18 year period (1965-1982) and analysed the data in three 6 year periods. Patients had been assessed using published prognostic criteria and the families counselled accordingly. If parents requested early surgical intervention, it was carried out regardless of the prognosis. They found that a good prognosis was given to more infants over the three periods (42% of 53 newborns, 1965-70; 58% of 65 newborns ,1971-6; and 71% of 94 newborns, 1977-82). Of the infants given a poor prognosis, more were treated with early surgery over the three periods (19% of 31, 1965-70; 33% of 27, 1971-6; & 52% of 27, 1977-82). Improved survival of the infants treated with early surgery was statistically significant over the three periods. They suggest a number of factors may have influenced the outcome. They are a referral hospital and selective referral may have influenced the outcome; social attitudes and technology changed over the period, in that

ventriculo-atrial shunts were used in the first period, ventriculo-peritoneal shunts in the second, and assessment of hydrocephalus had become non-invasive (using cranial sonography and computerised tomography) by the third period.

A retrospective review of patient records from Puerto Rico (1980-85) was published by Brau et al. (1990). They reviewed 128 case records and found that closure of the lesion occurred on average 6,6 days post delivery and that the incidence of ventriculitis in their population did not improve if the lesion was closed less than 48 hours post delivery. They found that the presence of post closure complications, such as skin flap necrosis, cerebrospinal fluid leaks and wound infection, was a statistically significant risk factor for the development of ventriculitis.

In an attempt to reduce the incidence of ventriculitis the use of prophylactic antibiotics has been tried. Kaplan (1981) suggested that in the early lesion (<24 hours), antibiotics should not be used routinely, but after 24 hours the lesion should be treated with saline swabs every 2 hours, and broad spectrum antibiotics should be administered intravenously for 24 hours before closure is undertaken. McLone et al. (1981) also suggested that effective antibiotics used for treatment and prophylaxis, especially against gram-negative organisms, is an important factor in the improved survival rate in myelomeningocele. Charney et al (1985) in their review of 110 patients having either early or delayed surgical closure of their myelomeningocele showed that although they found no association between the delay in surgery and the development of ventriculitis, antibiotic usage among infants having delayed surgery beyond 48 hours was associated with diminished likelihood of developing ventriculitis. However, Brau et al.(1990) found that the use of prophylactic antibiotics prior to closure of the myelomeningocele did not lower the incidence of nonshunt-related ventriculitis.

As surgical techniques and advances in the control of hydrocephalus have occurred over the last 30 years, so the survival rate has improved from about 50% in the earlier studies

(Sharrard et al., 1967), to over 80% in later studies (McLauglin et al., 1985). Most studies agree that early closure (<48 hours) improves survival and prognosis and reduces the number of infection related deaths. The use of prophylactic antibiotics in reducing the incidence of ventriculitis appears promising (Charney et al., 1985). The argument as to whether early closure improves lower limb muscle power as suggested by Sharrard et al. (1963) has not been substantiated by others (Brocklehurst et al., 1967; Smyth et al., 1974).

III.4.2 RESULTS OF UNSELECTED SERIES

A number of series of unselected newborns born with a myelomeningocele have been documented and some followed-up for a significant period. Lorber (1971) published his experience with an unselected population of 524 infants, all treated within the first day of life, 323 treated between 1959 and 1963, and 201 treated between 1967 and 1968. The first series were followed-up for 7 to 12 years, whereas the second series were observed for 2 to 4 years. Despite improved management between the first and second series which contributed to an improvement in survival from 50% to 64% in the two groups, he found that the proportions with serious intellectual or physical handicaps did not decrease in the second series. The infants who fared particularly badly were those with extensive paralysis at birth, those with a head circumference exceeding the 90th percentile by 2 cm. or more, and those born with a gross kyphosis or with other major congenital defects. This led him to draw up the initial selection criteria for surgical closure of the defect. These criteria were severe hydrocephalus at birth, total paraplegia, kyphosis, or any additional birth defect. A year later he published his experience of 270 consecutive unselected cases with criteria for selection (in addition to the above mentioned criteria he added thoracolumbar lesions), born between 1962 and 1964 (Lorber 1972). Two hundred of these patients were admitted and received active management on their first day of life, 31 were admitted between 2 days and 10 months of age, 3 had a skin covered myelocele and 36 a meningocele. The babies with one or more of his adverse criteria made up the majority of the deaths (40 of 63), were all severely physically

handicapped and half (30 of 62) had intelligence quotients below 80. Of the babies with no adverse criteria one third were moderately (10 of 42) and the remainder severely physically handicapped, and almost 38% (16 of 42) had intelligence quotients below 80. Compared to the original admissions half of those admitted without adverse criteria were of normal intelligence as compared with only one fifth of those with adverse criteria. However, if one looks at the survivors, 26 (62%) without adverse criteria and 32 (52%) with adverse criteria had normal intelligence, which statistically is not significant. Urinary incontinence was a major problem in both groups. This resulted in a publication of a detailed assessment of the neonate born with a myelomeningocele with specific reference to determining the sensori-motor level of spinal cord function in these infants (Stark, 1971).

Stark maintained a policy of selection of patients for operation because he found 40-45% of these infants were severely handicapped and unlikely to survive. The same year, Katzen published data from Johannesburg, on the outcome of 54 patients born between June 1962 and June 1970 with myelomeningocele treated within the first day of life. He showed a high mortality rate (55,5%), over half dying from sepsis and central nervous system infection within the first six weeks of life. The remainder died within the next two years from complications of shunt surgery (ventriculo-atrial shunts). The age at follow-up of the survivors varied from 8 months to 8 years. Of the survivors 15 (62,5%) had normal intelligence, 5 (20,8%) were educationally subnormal, and 4 (16,6%) were mentally retarded. In comparison to other studies at the time his survival rate was low (cf. Mawdsley & Rickham, 1969). He concluded that, unless complicated by another major congenital anomaly, active surgical intervention should be undertaken, as there was no way of predicting survival or the outcome at birth.

A year later, Ames and Schut published their experience of 171 children with myelomeningocele, who all underwent active surgical management. Thirty three (19%) died and twenty three (11%) were lost to follow-up. Of the infants followed-up 91 (79%) were ambulant, 70 (60%) were competitive with a developmental or intelligence quotient

>80 and 86 (76%) were incontinent. Of the 56 infants who were lost to follow-up or died, thirty had thoracolumbar lesions and would most likely have been non-ambulant and half of them non-competitive. However, they concluded that as there were no infallible criteria for predicting the outcome of these cases at birth, they advocated active surgical management to close the defect and relieve the hydrocephalus in all newborns with this condition.

Hunt et al. (1973) published their findings on 113 cases of myelomeningocele operated on within 48 hours of birth between July 1963 and January 1971. The eighty survivors were surveyed between March 1971 and March 1972. They analysed a number of clinical factors at birth for their predictive value with respect to outcome. Of these the sensory level (high - T5-T10, Intermediate T11-L3, Low L4-S5), which frequently differed from both the external and radiological levels of the lesion, correlated with the outcome in terms of mobility, intelligence, continence, major complications, and overall disability and also with deaths caused by renal failure. As the range of age at follow-up varied from 1 year 3 months to 7 years 8 months, not all survivors were assessed for all disabilities. Intelligence was based on an IQ obtained from the school medical service for those children attending school and a developmental assessment for the pre-schoolers (with no precise score given), and the child was placed in one of four categories (above average, average, below average or severely retarded). They concluded that the sensory level at birth could be used in combination with other selection criteria to predict the outcome in the newborn with myelomeningocele.

Stein et al. (1974) added to the selection criteria suggested for use in cases of myelomeningocele. They reviewed 163 children born with open myelomeningocele between 1963 and 1968, who had had a neonatal skull X-ray. At follow-up 151 93%) had survived, 75% received shunt surgery for hydrocephalus and 72% were ambulatory. They retrospectively grouped the infants into two groups, each adhering to Lorber's criteria for a poor and good outcome. Comparison of the two groups showed a significant difference only for ambulation, degree of handicap and intelligence. Regrouping the data

using the presence of craniolacunia or lacunar skull deformity on neonatal skull X-ray and at least two of Lorber's criteria as a predictor of a poor outcome made the comparison of the poor and good outcome groups highly significant for survival, requirement of shunt insertion, ambulation, intelligence and the degree of handicap. They recommended the use of neonatal skull X-ray and Lorber's criteria in the process of selection of patients with myelomeningocele for early surgery. Their follow-up group excluded patients without a skull X-ray, which could have been because of early demise or not indicated as hydrocephalus was absent. The high survival rate (93%) is suggestive of a selected population. Lorber (1975) claimed that his experience using craniolacunia on a similar number of infants born between 1967 and 1969, could not confirm these findings. Hunt and Holmes (1976) in their analysis of 83 survivors of their consecutive series of 116 children with spina bifida cystica, found that the need for a ventriculo-peritoneal shunt was significantly related to the presence of craniolacunae on the neonatal skull radiograph.

Soare and Raimondi (1977) published their series of 173 children with myelomeningocele. Their inclusion criteria was survival to the time of testing and therefore nonsequential and they assessed the children's intellectual and perceptual-motor skills. The majority of the children developed hydrocephalus and required shunt surgery (133), and 63% of these children were shown to have intelligence quotients above 80, compared to 87% of the children without hydrocephalus. They compared the children to their siblings and found that those with hydrocephalus were less intelligent than their siblings, whereas those without hydrocephalus were not. When matched for age and IQ with siblings, the patients fared less well on perceptual-motor functioning testing, as did the patient group with hydrocephalus compared to the patients without hydrocephalus. Brown and McLone at the VIIth Congress of the European Society of Paediatric Neurosurgery reviewed the medical records of 167 of the original 173 patients in Soare and Raimondi's study and regrouped the patients, placing them in one of three categories: 1. not shunted (39), 2. shunted without CNS infection (86), and 3. shunted with CNS infection (42). No patient without a shunt sustained a CNS infection. The mean intelligence quotients for those not requiring a shunt and those who were shunted but remained infection free were within the normal range, except that those

requiring a shunt had lower scores (102 vs 95). Those children with CNS infection complicated shunting had a significantly lower mean intelligence quotient (73). The cerebrospinal fluid analysis and the duration of the infection could not be correlated with the intelligence quotients. They concluded that the result of infection on the developing CNS is a significant factor in reducing the intelligence quotient in myelomeningocele. The data were reported in 1981 and published a year later (Brown and McLone, 1981; McLone et al., 1982).

Naglo and Hellstrom (1976) followed 59 unselected patients born between 1964 and 1971 with myelomeningocele and closed on the first day of life. They were followed up in 1973-4 by which time 11 had died. Despite the small numbers involved, at follow-up the physical handicap was similar to that seen in Lorber's study on unselected patients but the rate of moderate to severe mental handicap was lower (16% versus 36%) (Lorber, 1971), and compared favourably with results from a selected series (Stark and Drummond, 1973). It is suggested that broader indications for shunt insertion, closer shunt supervision and liberal indications for shunt revisions were responsible for the reduced incidence of mental handicap in this series.

Lorber (1975) published further follow-up on 848 infants born between 1959 and 1968. Despite active management only 50% survived, most dying in the first year of life. Later deaths were shown to occur at about 2% per annum and the commonest cause of these later deaths being shunt complications and progressive renal disease. Only 6 of the survivors (1,4%) were free of handicaps and a further 73 (17,2%) had moderate handicaps. Three hundred and forty five (>80%) of the survivors had severe multi-system physical defects and 201 (59%) had normal intellectual abilities, 93 (27%) were moderately retarded and 51 (14%) were severely retarded.

Lister et al. (1977) reported on 200 unselected consecutive closures of myelomeningoceles during 1962-1964, all closed within 36 hours of birth, the majority within 24 hours of birth (193). The survivors were reviewed at five and ten years of age. Eighty three of

the children died within the first five years, the majority within the first three months of life, usually as a result of intracranial haemorrhage or infection. The older children died of either shunt or renal complications. Of the survivors 117 were seen at 5 years of age. The majority of the survivors had a normal intelligence (80), with only one being profoundly retarded. Ninety nine were ambulant (67 with calipers), 24 were continent and a further 51 were 'acceptably dry'. At 10 years of age 106 survivors were seen, 5 having died and 6 were lost to follow-up. Intelligence quotients remained unchanged, the number continent had increased (26), a further 39 were 'acceptably dry', and the number wheelchair bound had risen to 36 (38%) with a number of others spending a great deal of their time in wheelchairs. Forty seven of survivors seen at ten years of age had a 'poor' grading, with 59 (56%) having a 'good' grading, taking their intelligence, continence and ambulating into account.

McLone et al. (1981) from Chicago reported on 100 unselected consecutive cases treated for myelomeningocele from 1975 to 1978. The majority (91%) were closed within 24 hours of birth, 74 developed hydrocephalus requiring shunt surgery, and the overall mortality was 6% (due mainly to infection). Following surgery at 10 days of life a repeat motor examination showed a significant improvement in motor function, a finding also noted in unoperated cases. The follow-up interval varied from only 2 months to 4 years and comments were therefore restricted to mortality, complications of management and infection in this group. A further follow-up on these patients was published in 1983 (McLone 1983). The follow-up period was then from 3,5 years to 7 years. The overall mortality was then 14%. Of the children requiring shunt surgery 52% required a shunt revision (16% requiring only one revision). Of the survivors 73% had developed normally and 54% were community ambulators (walks indoors and outdoors, may need crutches or braces or both, and use a wheel chair only for long outings) and only 8% were wheelchair bound. Of the 86 survivors over four and a half years of age 87% had achieved urinary continence, mainly on clean intermittent catheterisation in combination with pharmacological agents. Of these survivors 47% were intellectually normal, were continent of urine and were community ambulators. An additional 28% were intellectually normal and

continent, but were not community ambulators. By 1985, a further 100 cases had been included in the study (McLone et al., 1985; McLone, 1986). By this stage ambulation was approaching 75% for the entire group. They reported that although 73% of the survivors had an intelligence quotient above 80, eye-hand co-ordination was noted to be diminished in many, and learning disabilities were coming to the fore. This study of unselected cases compared favourably with the results Lorber obtained in his selected group of patients (Lorber & Salfield, 1981). In comparison with the Lorber study the mortality for the actively managed patients was almost identical, more required shunt surgery, fewer (15%) had normal intelligence, and more were continent and ambulant.

Not all patients with delayed closure of the lesion do badly. Okorie et al. (1987) from Sheffield reported on 13 cases of delayed closure and compared them to 22 cases of early closure from 1980-1985. The intelligence quotients in both groups were similar with more infants requiring shunting in the group having delayed closure.

Hunt (1990) reported further on their cohort operated on from 1963 and 1970, pointing out the wide range of disabilities seen in patients with myelomeningocele. Sixty nine of the original 117 patients had survived. Sixty were shunted and 2 were blind following shunt dysfunction. Forty seven had an IQ >80, 35 were wheel chair dependant, 52 were incontinent, 33 were unable to live independently without help or supervision and only 17 were in open employment. A 25 year follow-up on these patients in 1992 (Hunt and Poulton, 1995) reflected similar findings. Sixty one patients were still alive, the lowest survival being in children with a sensory loss extending above T11. Forty three (70%) with a normal IQ, and 20 were community ambulators. Sixteen (26%) were fully continent, the remainder using a variety of methods to achieve continence, in 14 of whom management was still unsatisfactory. Thirty three were living independently, only 11 now requiring supervision and help. Seventeen needed daily care which was provided by the parents. They found that the two main determinants of disability were the extent of the neurological deficit and the IQ. Long term follow-up studies are extremely

valuable and reflect the outcome of a variety of treatment modalities especially in the management of hydrocephalus and urinary incontinence.

Since Lorber suggested his selection criteria, the results of unselected series have shown an improvement in survival and outcome as the surgical, urological and medical care have improved. The results, as reported by McLone et al. (1985) are similar to those of selected series (Lorber and Salfield, 1981). Questions regarding Lorber's criteria have arisen. McLone et al. (1981) reported that the motor function in lower limbs could improve up to 10 days post delivery, with or without surgery, bringing the Lorber criterion of paraplegia at birth into question. They also found that poor intellectual development was linked to CNS infection and not necessarily only hydrocephalus, which questions the role of hydrocephalus as a selection criterion (McLone et al., 1982). The multiple handicaps seen in myelomeningocele have been well documented in a 25 year follow-up study and the role played by the extent of the neurological deficit and IQ in determining disability and dependency emphasised (Hunt and Poulton, 1995).

III.4.3 RESULTS OF SELECTED SERIES

Stark and Drummond (1973) published data on 163 infants who were admitted to their unit in Edinburgh between 1965 and 1971. Although unselected prior to admission, they were then divided into two groups, one with good prognostic criteria (78) who received early surgical intervention, and the other (85) with poor prognostic criteria who received conservative management only. The criteria were similar to those suggested by Lorber (Lorber, 1971) and included severe paralysis, gross head enlargement and spinal deformity such as kyphosis. Over 80% of those treated conservatively were dead within 3 months of life and less than 10% survived to 6 years of age, whereas although 18% of those managed actively died within 3 months of life, 70% survived to 6 years of age. The outcome was assessed and compared to Lorber's data. Overall survival was 40%,

which is lower than of Lorber's study (60%), but the survivors were less severely handicapped. Only 25% were not ambulant (cf. 49%), 16% had upper urinary tract damage and incontinence (cf. 34%), and only 20% had a combination of mental and severe physical handicap (cf. 36%). The most severe handicaps were associated with complications, notably central nervous system infection. They suggested that selection offered the best prospects of independence in spina bifida.

In the same year, Lorber (1973) reported on 37 babies subjected to his selection criteria. Of the 37, 25 fulfilled one or more of his criteria and were expected to have a poor prognosis for quality of life. These babies were not treated and all died within 9 months. This report was updated when Lorber and Salfield (1981) published the results of their selective treatment in 120 infants with open spina bifida, which included the original 37 babies previously reported. The patients were admitted between 1971 and 1976. Seventy one of the infants had adverse criteria and were not treated, all dying, 90% within 6 months of birth. The remainder were all treated. Seven had meningocele, all of whom survived free of handicap and 42 with myelomeningocele, 36 of whom survived and were followed-up after 3 to 9 years. At follow-up, which varied from 3 to 9 years, only 25 of the survivors had formal intelligence quotient assessments and those not assessed were said to be 'normal', 33 were ambulant (11 with aids), and 30 had normal upper urinary tracts, of whom 20 were continent. Despite the selection process 8 children (22%) had moderate to severe handicap.

Shurtleff et al. (1974) presented their data on the fate of 371 patients with myelodysplasia, including 88 patients who had received supportive care. Their criteria for selection differed from Lorber's in that aggressive management was recommended for any infant, regardless of the level of the lesion, if a good intellectual prognosis was noted. Their other criteria included a frontal cerebral mantle >1,0 cm or brain mass >60% of the mean for age, absence of a systemic disorder that could cause hydrocephalus and severe brain malfunction, absence of radiological and neurological signs of severe brain deformity, absence of noteworthy CNS bleeding and infection, absence of major

malformations that would preclude self-care as an adult, and a family with economic and intellectual resources who lived within reach of an appropriate medical facility or a commitment by a social agency to provide needed resources for medical care costs. They emphasised that a complete discussion with the family had to be included as part of the process. Of the patients given supportive care only, 52 were born before 1965, when an evaluative process of the newborn was begun, and 36 were born after 1965. Of the former group, 30% survived to the second decade and 10% of the latter group survived the second year of life, demonstrating that some of these infants survived despite being given a poor prognosis.

Gross et al. (1983) from Oklahoma attempted to evaluate a programme for the early management and decision making for the treatment of myelomeningocele. They recommended supportive care for infants with the published adverse criteria and used a team approach to come to the decision and to counsel the parents. They evaluated 69 infants from 1977 to 1982. Thirty six (52%) received vigorous treatment, whereas for 33 (48%) only supportive care was recommended, which was accepted by 28 families. The other five families requested early vigorous treatment, 3 of the babies surviving. Of the babies in supportive care, 24 died (mean 37 days), 3 were treated later at their parents request and one had moved and no follow-up was available. The supportive care was provided in a children's shelter (an intermediate nursing facility). The authors suggested that, using a co-ordinated approach, those receiving supportive therapy only would die and die quickly, which was not a universal finding. Unfortunately, they provide no death data on these infants, nor did they document how the survivors of the no treatment group had fared. Their ethical approach was quite severely criticised by Freeman in 1984.

Kaiser and Rudeberg (1986) published data from Switzerland. They studied 46 children from 1977 to 1984 with myelomeningocele. Using the presence of three of Lorber's criteria, 8 (17%) received no active management and the remainder were operated on within 24 hours. Twelve of the actively managed group had two or less adverse criteria,

required more shunt surgery than the rest, were less ambulant but were intellectually no different from the others managed actively. Of the eight not managed actively, five died within the two months, the remainder requiring delayed shunt surgery and back closure. Thus despite using three of the published Lorber criteria, three of the eight infants fulfilling the criteria survived. They suggested that at least three of the adverse criteria, especially the three last ones (hydrocephalus, birth injury and other major congenital defects) and social factors, should be considered when selecting infants with myelomeningocele for surgery.

In a retrospective folder review of 110 consecutive neonates referred with myelomeningocele from 1978-1982, Charney et al. (1985) and Sutton et al. (1986) from Philadelphia looked at the question of selection. The parents of the infants were given an option of early surgery or conservative medical management when presented with the possible prognosis regarding ambulation, sphincter continence, hydrocephalus and general survival, with and without surgery. Fifty two (47%) were closed within 48 hours of birth, 41 of whom had no adverse criteria. Thirty two (29%) were closed 3 to 7 days after birth, 18 of whom had no adverse criteria. Twelve (11%) were closed a week or more after birth, all with adverse criteria. Fourteen (13%) did not have surgery, all of whom had adverse criteria and died by 10 months of age, the majority within the first 8 weeks. Of the infants who received surgical management, 92% of those operated on within 48 hours of birth, 94% operated on from 3 to 7 days of age, and 100% of those operated on one week or more after birth survived. Ten infants operated on developed ventriculitis, but there was an association with the timing of surgery. Developmental delay (DQ<80) was present in 37% of the infants and was related to the severity of the lesion at birth, but not the timing of the surgery or the presence of ventriculitis acquired because of the delay in surgery. The study, although limited being a folder review and having a variable follow-up period, did show a lower mortality than other studies (Gross et al., 1983; Lorber 1973) suggesting that more infants are operated on if the parents are empowered and fully involved in the decision making process.

Katzen (1981) reported on the application of Lorber's selection criteria to 79 infants born with a myelomeningocele at the Johannesburg Hospital. Thirty four were untreated of whom 27 died within the first month of life, a further 2 within a year, one at eighteen months, one at four years and one at six years of age. Two had survived beyond six years of age. Thirty of the 45 treated infants survived (66%), and were considered to be only moderately handicapped, details of which are not reported.

Collis (1972) highlighted the fact that not only were selection criteria in myelomeningocele a problem for medical staff, but they also impacted on the nursing staff. He showed that using these criteria, the survival of infants admitted to their neonatal unit with myelomeningocele had dropped from 90% in 1967 to 50% in 1972. Although this created space for other admissions, the nursing staff had difficulty with implementation, especially the idea of not feeding the infants who, because of a poor prognosis, were not for active surgical intervention.

Not all patients denied active surgical management for their myelomeningocele die. Guiney et al. (1984) found that one third of patients with an open myelomeningocele at birth treated conservatively initially had a normal IQ, although not all these children underwent delayed back closure (Guiney et al., 1984; Guiney et al., 1986). Menzies et al. (1985) from Newcastle upon Tyne found that 8 of 27 children not offered early surgery but family centred care survived to school entry. All were chairbound and incontinent, but none was intellectually retarded.

Selected series initially showed improved outcome in children with myelomeningocele but at a price. If those selected for no surgical intervention are included in the mortality, the mortality rates are high (60%), which is higher than mortality rates achieved in the 1960's without selection. However, the intellectual outcome reported for most selected series is approximately 15% better than for unselected series for IQ's within the normal range (Lorber and Salfield, 1981; McLone et al., 1985). Whether this marginal increase in

intelligence is worth the increase in mortality is questionable. Selection also impacts on families who are frequently asked to help in the decision making process, and the nursing staff who have to nurse the untreated patients through difficult days until their demise (Collis, 1972). With advances and improved outcome reported from more recent unselected series, it is difficult to justify selection purely on the grounds of an improved outcome (McLone et al., 1985).

III.4.4 PREDICTABILITY

Laurence et al. (1976) pointed out the difficulty in predicting the outcome reliably in spina bifida. The clinical findings in 85 cases with spina bifida (6 meningoceles, 4 encephaloceles, 71 open myeloceles and 4 closed myeloceles) were given to two neurosurgeons and two paediatricians, who were asked to predict from them the length of survival and quality of survival with regard to intellect, locomotion and continence. Forty four of the children were born between 1964 and 1966 and had received active surgical management and 41 were born between 1971 and 1974 and received conservative management. The clinicians all correctly predicted the outcome in the case of the meningoceles, encephaloceles and the closed myeloceles. However, in the case of the open myeloceles, the paediatricians correctly predicted the survivors, but included some who had died; whereas the surgeons correctly predicted the deaths, but included some of the survivors. All correctly predicted limb and sphincter functions in the survivors. All underestimated the intellect of the patients not requiring shunt surgery and those with successfully shunted hydrocephalus, and overestimated the intellect of the patients who had developed intracranial infection and shunt blockage. This study helped point out the difficulty in predicting outcome at birth, and the inherent dangers of a policy of selection which depends on the ability to predict outcome at that early stage.

A decade later Siperstein et al. (1988) looked at paediatricians prognostications in cases of myelomeningocele. They asked 604 paediatricians to complete a questionnaire on 3

hypothetical cases of newborn infants with myelomeningocele (one without hydrocephalus, one with moderate hydrocephalus and one with severe hydrocephalus). Only 373 (62%) returned their questionnaires. The paediatricians prognostications were least optimistic for the case with severe hydrocephalus, and most optimistic for the child with no hydrocephalus. This affected how medical information would be presented to the parents, whether or not they would comply with the parents' desire not to undertake surgery and what they would do if the child were their own. They were not influenced by the level of the lesion and the severity of paraplegia in their prognosis. This study suggested that paediatricians were being influenced by the presence of hydrocephalus, despite the fact that it is difficult to predict the intellectual capabilities in myelomeningocele with associated hydrocephalus.

Predicting outcome in the case of myelomeningocele is not an easy task, as has been shown by the two studies reviewed (Laurence et al., 1976; Siperstein et al., 1988). Intellectual outcome appears to be the area which influenced predictions more than future limb paralysis. However, it proved to be difficult to predict, especially when associated with hydrocephalus.

III.4.5 OTHER FACTORS

Quinn and Boston (1987) investigated parental attitudes to the unclosed open neural tube defect. The majority had no difficulty dressing the lesion and found that the child was more comfortable, easier to handle, and more socially acceptable after closure of the lesion.

Delight and Goodall (1988) reviewed 98 babies born from 1971 to 1981 and considered not suitable for surgery. Sixty three of these infants survived more than one week and 44 of the families were interviewed five to fourteen years later (9 could not be traced and the

remainder refused to be interviewed). Twenty six of the babies had remained in hospital until death, whereas the remainder were allowed to be taken home. The babies taken home lived longer than those left in hospital. The parents of those left in hospital were sadder and considered the life of their baby to have been one of poor quality, whereas those who had taken their babies home had a more positive view of their child's life.

Haynes et al. (1974) published their study looking at the effect on socio-economic status on the outcome of patients with myelomeningocele, suggesting that the risk of infection and mortality might be much higher for the indigent patient as compared to the nonindigent patient with myelomeningocele. They reviewed the outcome of infants born with myelomeningocele between 1964 - 1971 and managed at a private and a public hospital. Thirty eight patients were treated at the public hospital (indigent patients) and 52 patients were treated at the private hospital (nonindigent patients). They showed a 50% mortality in the indigent group versus a 22% mortality in the nonindigent group. Approximately sixty percent of both groups required shunting for hydrocephalus. Forty two percent of the indigent group and 31% of the nonindigent group were lost to follow-up. Both groups had similar non-shunt related infections, but 44% of the indigent group had shunt-related infections versus only 15% of the nonindigent group. No details of delays in initial surgery are given for either group and the loss to follow-up is too high to draw any valid conclusions. This may also reflect the hospital environment and level of nosocomial infection in the hospitals studied and have little to do with the socio-economic status of the affected families.

Views regarding children with myelomeningocele differ from the western view in African culture. Oyewole et al. (1985) carried out a prospective trial in Ibadan from 1982 to 1983. Seventy six new patients with myelomeningocele were studied and their parents interviewed. Of the 76 couples, 74 had neither seen or heard of the condition. The majority had no idea of the causation and one-third incriminated witchcraft. Their main concern was the lesion itself, the better educated parents demanding surgical management. Twenty two of the patients were managed surgically. As no intervention was offered to

the majority of patients, the default rate was high. It is suggested that many of the defaulting children died from infections. In a number of cases once the lesion could no longer be hidden, the families used various means to get rid of the malformed child. Most mothers showed relief rather than remorse on the death of their child.

The role of parents and a home environment are important in the care of any infant. Most parents care better for the children than anyone else and the level of infection in a home environment is far less than that in hospital and children selected out for conservative care only, may well live far longer in their own home environment and allow the parents time to bond, care and eventually mourn for their baby more successfully (Delight and Goodall, 1988). The role of the family's socio-economic status in the survival and outcome of infants with myelomeningocele has been questioned (Haynes et al., 1974). Very few studies have looked at cross-cultural customs, traditions and feelings with regard to a baby with a myelomeningocele. In Africa, we are aware of these differences and the way western medicine may be in conflict with these beliefs and views is important to understand (Oyewole et al., 1985).

III.4.6 HYDROCEPHALUS MANAGEMENT

In patients with myelomeningocele, ventriculomegaly is common at birth even without overt hydrocephalus. The majority of the infants with myelomeningocele will develop hydrocephalus, but this rarely becomes manifest until after surgical closure of the myelomeningocele. In an attempt to identify those infants with ventriculomegaly who were likely to develop hydrocephalus, Bell et al. (1987b) measured the lateral ventricular ratio (maximal ventricular width/maximal hemispheric width) using cranial sonography, and defined ventriculomegaly as a lateral ventricular index of greater than 0,32. They monitored 25 neonates, and found a lateral ventricular ratio greater than 0,32 in 23 of the neonates. All but one of these infants required a ventriculo-peritoneal shunt for

hydrocephalus. The 2 with normal ratios did not develop hydrocephalus. They suggested that the cerebrospinal fluid may escape by draining down the patent central canal of the spinal cord and the exposed neural placode, which would explain the rapid progression of hydrocephalus following closure of the back in some cases. The presence of various brain anomalies with myelomeningocele is well known, particularly the Chiari II malformation. Its association with hydrocephalus is important. Most centres utilise either CT (Computerised Tomography) or MRI (Magnetic Resonance Imaging) scanners but these are expensive and often not easily accessible. De la Cruz et al. (1989) from Madrid reviewed 25 children with myelomeningocele over a five year period who were evaluated using cranial sonography. Twenty three of the cases had a Chiari II malformation with a 'bat-wing' configuration of the frontal horns and a downward displacement of the cerebellum, obliterated basal cisterns and low positioning of the tentorium cerebelli. Hydrocephalus was easily seen and monitored prior to shunt surgery and following it. They suggested this as convenient and inexpensive way of assessing the brain anomalies in children with myelomeningocele.

Although ventricular shunting has revolutionised the care of children with hydrocephalus, problems with shunts are common. Most studies in this area have grouped children with hydrocephalus together, regardless of aetiology (Riva et al., 1994). Liptak et al. (1985) documented shunt problems in 67 children with a neural tube defect and hydrocephalus. Twenty eight percent of the shunts failed within the first 6 months after insertion, 37% within the first year and 50% by four and a half years after insertion. Shunts inserted in the first year of life were much more likely to fail than those inserted after 1 year.

Ten cases of the early repair of myelomeningocele and simultaneous insertion of a ventriculo-peritoneal shunt (from 1975 to 1985) was reported by Hubballah and Hoffman (1987) from Toronto. They had one fatality in the post operative period secondary to aspiration. The remaining nine patients were followed-up at different stages, from 1 to 9 years of age. No shunt infections were reported, but a number of shunt revisions were required. Neuropsychologically, 3 of the children were normal, 4 were borderline, and 2 were grossly retarded. They unfortunately made no comment on hospital stay and no

comparison with children with myelomeningocele who had delayed placement of a ventriculo-peritoneal shunt for hydrocephalus. That same year, Bell et al. (1987a) from New York published their results on 17 patients who had a one-stage myelomeningocele closure and ventriculo-peritoneal shunt placement. They had 1 shunt infection and found that there was little correlation between ventriculomegaly on preoperative computed tomography and head circumference. Again there was no comment on hospital stay. Chadduck and Reding (1988) from Arkansas, studied 22 patients who had simultaneous shunt placements and repairs and 11 patients who had shunt placements 6 to 14 days after the repair. There was no operative mortality nor were there infections within 30 days of surgery, and hospital stay was similar in both groups. There were single infections later in both groups, and fewer shunt revisions in the simultaneous shunt placement and closure group. It is suggested that in the face of ventriculomegaly in myelomeningocele, simultaneous shunt placement at the time of repair is a safe and viable option. Although hospital stay is not shortened, the child is spared a second operation and anaesthetic.

The majority of children with myelomeningocele have hydrocephalus and will require a shunt procedure (Bell et al., 1987a). Ventriculo-peritoneal shunts are the preferred procedure, but some authors suggest that simultaneous shunt placement with lesion closure save the child a second operation and anaesthetic (Hubballah and Hoffman, 1987; Bell et al., 1987a; Chadduck and Redding, 1988). However, not all children with myelomeningocele will develop hydrocephalus requiring shunting and not all have evident hydrocephalus at the time of lesion closure, especially if it is done early, and by waiting the child may be spared an unnecessary shunt procedure. The rate of shunt failure appears to be greater in those shunts placed in the first year of life and in view of the dangers and complications of shunt failure surveillance should be high during this period (Liptak et al., 1985). Cranial sonography is now considered an easy noninvasive method of diagnosing and monitoring hydrocephalus, while the anterior fontanelle remains patent.

III.4.7 INTELLECTUAL PROBLEMS

Badell-Ribera et al. (1966) analysed the data on 75 patients between the ages of 5 and 21 years. In sixty two of the patients with a history of rapid head enlargement and no neurosurgical treatment the hydrocephalus eventually became non-progressive. This hydrocephalic group scored lower in psychological testing and presented a significant discrepancy between verbal and performance scores which they considered a sign of brain damage. They further divided the patients into 5 groups on the basis of their functional disability. The IQ (Intelligence Quotient) score increased with the decreasing degree of physical disability in the groups, but was related to the presence of hydrocephalus. IQ scores of the patients with the same severe paralytic defect but without hydrocephalus were normal. They concluded that to improve the prognosis of these children early and effective treatment of their hydrocephalus is important.

Beks et al. (1966), in their review of 162 children with spina bifida, of whom 98 were followed-up, suggested that the intellectual development of children with spina bifida is practically normal, provided the hydrocephalus which may develop can be treated early and successfully, however, only 37 were tested psychologically (20 who had hydrocephalus). Their findings were optimistic, but no differentiation was made on the outcome of the children with meningocele, and only a small sample of the children underwent formal psychological testing.

In a small series Scherzer and Gardner (1971) studied 14 surviving children with myelomeningocele. Three did not have surgery, the others had surgery ranging from the first day post delivery to beyond 1 year of age. Eight children had normal IQ's, 3 were mildly and 3 moderately retarded. Hydrocephalus developed in 9 children and spontaneously arrested in all but 1 child. Five of the children were of normal intelligence and hydrocephalus specifically was not a meaningful predictive factor for this group. Intellectual potential correlated with social class.

Heimburger's (1972) review of 104 patients followed-up at age 13 to 19 years, showed that 28 of the 37 survivors, all of whom had early closure of their lesion, were attending school, 22 in age appropriate classes. Of the nine not attending school, 4 were mentally retarded.

Lorber (1972) in his follow-up of 270 unselected patients with myelomeningocele with criteria for selection, showed that compared to the original admissions half of those admitted without adverse criteria had normal intelligence as compared with only one fifth of those with adverse criteria. However, of the survivors, 26 (62%) without adverse criteria and 32 (52%) with adverse criteria had normal intelligence, which was not statistically significant.

Katzen (1971) showed that of 54 patients born with myelomeningocele treated within the first day of life, and the survivors followed-up at 8 months to 8 years of age, 15 (62,5%) had normal intelligence, 5 (20,8%) were educationally subnormal, and 4 (16,6%) were mentally retarded.

Spain (1974) assessed the verbal and performance abilities of 129 three year-olds with spina bifida and 16 with encephalocele, using the Hand-Eye Co-ordination and Performance Scales of the Griffiths Mental Development Test and the Reynell Developmental Language Scale. Seventy three (58%) of the patients with spina bifida had a developmental quotient <80. Ninety six children requiring a shunt to control their hydrocephalus, showed lower mean scores on all tests than children without shunts, particularly in the non-verbal tests. Only one third of the children with shunts seemed to be developing normally. The presence of a shunt was also associated with moderate or severe physical handicap. Children with shunts and with below-average performance scores also tended to show poor verbal scores, and 40% were rated as hyper-verbal. They all showed good syntax but poor comprehension and inability to use language creatively on the verbal scales.

Hunt and Holmes (1976) analysed the results of 83 survivors of a consecutive series of 116 children with spina bifida cystica operated on within 48 hours of birth, from 1963 to 1971. Age at follow-up ranged from 13 months to 7 years 8 months. Intelligence quotients were taken from the school medical or hospital service records for school going children and the pre-schoolers were assessed using a developmental assessment. The results were grouped into either normal or subnormal. Of those children with a central nervous system infection, intelligence was impaired in 7, 6 being severely retarded. In the 9 children who did not suffer a central nervous system infection or require a shunt, intelligence was normal. In the 67 children who did not suffer a central nervous system infection but did require a shunt, intelligence was related to the sensory level found at birth and to the thickness of the pallium measured within 4 weeks of birth, and was unrelated to their head circumference at birth, or to its increase before shunt insertion, or to the number of shunt revisions required.

Soare and Raimondi (1977) using a wide range of psychological tests, studied 173 children with myelomeningocele of whom 133 had hydrocephalus with a shunt. Eighty siblings acted as controls. Children with associated hydrocephalus were found to be significantly less intelligent than their siblings or those without hydrocephalus. When matched for age and IQ, patients scored significantly lower than their siblings on a perceptual-motor functioning test, those with hydrocephalus lower than those without hydrocephalus.

McLone et al. (1982) compared the intellectual development of 167 patients with myelomeningocele in three groups - nonshunted, shunted and those with a history of ventriculitis. The mean IQs for the nonshunted group was 102, for the shunted group 95 and those shunted with a history of ventriculitis 72. A similar trend was found for visual motor integration. These results indicated the important role CNS infection plays and the non predictability of uncomplicated hydrocephalus in the intellectual development of infants with myelomeningocele.

Mapstone et al. (1984) reviewed 75 infants with myelomeningocele. They found a mean IQ of 104 for infants not requiring shunt surgery, 91 for those shunted without complications, and 70 for those shunted who had complications. These were significant differences and not explained by difference in spinal lesion levels.

Tew and Laurence (1984) studied 51 survivors of the 121 largely unselected live-born spina bifida children born in South Wales between 1964 and 1966. They and their controls were given an individual psychological assessment within a month of reaching their 16th birthday. The mean IQ of children with spina bifida was significantly lower (26 points) than their controls and were below the normal range. Only 16% of the spina bifida children had an IQ >100 (cf. control 51%) and 43% had an IQ <70 (cf. control 7%). They consequently had greater problems passing examinations and coping with reading and mathematical skills. The requirement of a shunt for hydrocephalus and the presence of severe handicap (graded according to locomotor deficit and urinary incontinence) were factors negatively affecting academic achievement.

Shaffer et al. (1985) examined IQ and achievement status of 60 children with myelomeningocele who had either uncomplicated hydrocephalus (shunted but without CNS infection or bleeds) or no hydrocephalus. In examining the effects of both hydrocephalus and functional motor level, they found an inverse relationship between IQ and functional motor level in children without hydrocephalus but not in children with hydrocephalus.

Wills et al. (1990) looked at intelligence and achievement test scores for 89 children with myelomeningocele, ranging in age from 4 to 14 years. The scores were obtained from the medical records which were performed at routine outpatient visits and excluded patients (11) with any history of ventriculitis. The patients with myelomeningocele scored lower on virtually all tests administered, but had particular difficulty with Performance IQ subtests, the Visual Motor Integration (Beery) and arithmetic calculation. With increasing age they found that these children fell further behind their peers in these areas but kept

pace on reading and spelling tests. They suggest their findings may reflect a developmental difference between children with myelomeningocele and non handicapped children in a visual-perceptual-organisational cognitive function.

Friedrich et al. (1991), from the same institute as Shaffer et al. (1985), used the same inclusion criteria and selected 73 children with uncomplicated myelomeningocele who were aged six and a half to sixteen years of age. The overall level of academic achievement and measured intelligence was similar to that previously reported by Shaffer et al. (1985). Defects in visual motor abilities and computational arithmetic were similar to that reported by Wills et al. (1990). The inverse relationship between the functional motor level and the presence of hydrocephalus reported by Shaffer et al. (1985) was only weakly supported. However, both studies used small samples which limited the detection of real differences.

Casari and Fantino (1992) looked at the effect of CNS infection and intellectual development in children with myelomeningocele. They reviewed 115 patients with myelomeningocele, 106 developing hydrocephalus and 92 of whom required ventriculo-peritoneal shunting. Twenty seven of these shunted patients developed shunt complications, 12 of whom were infected. They ranged in age from 2 to 11 years, with a mean age of 5 years. Nine of the patients with infected shunts were psychologically evaluated. Their IQ's ranged from 23 to 81, and were not significantly different from those of uncomplicated shunted patients with myelomeningocele (42) or those not requiring a shunt procedure (14). No relationship could be shown between the extent of infection or the number of shunt revisions and the IQ score.

The presence of hydrocephalus and CNS infection appear to be the two major determinants of intellectual outcome in myelomeningocele, the latter being the more important (Hunt and Holmes, 1976; McLone et al., 1982; Casari and Fantino, 1992). It is also accepted that the absence of hydrocephalus in myelomeningocele allows for a better

prognosis, but compared to their siblings, although functioning in the normal range for intelligence, there are suggestions that they do not do as well. Some of the earlier studies were optimistic about intellectual outcome, especially those using variable follow-up periods, using different assessments in the various age groups which may have led to the optimism (Beks et al., 1966; Heimberger, 1972).

III.4.8 ORTHOPAEDIC IMPAIRMENT

Because of the paralysis in the lower limbs of patients with myelomeningocele, large numbers of these children require orthopaedic treatment. Two commonly encountered congenital deformations requiring surgery are talipes equinovarus and dislocation of the hip.

Talipes equinovarus deformities may present secondary to immobile feet deformed by intrauterine pressure or to imbalanced muscle forces in the feet and ankles in the child with a low-lumbar/sacral level lesion. In addition the following features may be present - in the foot showing preservation of L4, there is often a contracture holding the foot in dorsiflexion and inversion, with the preservation of L5, a calcaneal foot deformity results, and if S1 is preserved the result is a cavus foot. Initial management involves serial casting, usually started in the newborn period, but subsequent surgical correction and the use of braces is frequently required to maintain alignment (Stark, 1971; Liptak et al., 1988).

Paralysis of the abductor and extensor muscles of the hip will produce a flexion-adduction deformity and, in a high proportion of cases, dislocation of the hip, if active power remains in the flexor and adductor muscles of the hip (Starch, 1967; Carroll and Sharrard, 1972). Hip instability in myelomeningocele is thus usually associated with mid-lumbar/upper lumbar/thoracic level neurological lesions. In lesions above L3, who have no potential to walk, relocation of dislocated hips are not carried out as they will derive no benefit from the procedure (Asher & Olson, 1983; Fraser et al., 1992). Carroll and Sharrard (1972) described

the natural history of hip instability in myelomeningocele, and reported a 65% of hip instability at birth and 76,6% at two to three years. In a review of 53 patients born with myelomeningocele 5 to 10 years following hip surgery, they found 23 remained unstable. They also documented a higher incidence of hip instability in patients with innervation to the 3rd/4th lumbar segment (28 of 43) than that of those with innervation to the 1st/2nd lumbar segment (9 of 17). There was a lower incidence of hip instability at birth in the patient with innervation to the 5th lumbar segment (3 of 6). None of the hips in patients with innervation to the first sacral segment, and only one of five hips in patients with innervation to the 12th thoracic segment was unstable.

Hoffer et al. (1973) suggested dividing patients with myelomeningocele into four categories: community ambulators (walks indoors and outdoors, may need crutches or braces or both, and use a wheel chair only for long outings); household ambulators (walks only indoors, may need a wheel chair for some indoor activity); non-functional ambulators (walking for therapy only, otherwise use wheel chair); non-ambulators (wheel chair bound). In a small select group of 56 patients with myelomeningocele, followed for 5 years, they found that all patients with thoracic neurological levels were non-ambulators. All patients with sacral lesions were community ambulators. Of the 40 patients with lumbar neurological levels, 14 were community ambulators (6 upper and 8 lower lumbar lesions), 5 were household ambulators (3 upper and 2 lower lumbar lesions), 2 were non-functional ambulators (both lower lumbar lesions), and 19 were non-ambulators (10 upper and 9 lower lumbar lesions). They suggested that lower limb surgery be generally reserved for potential functional ambulators and that it be performed prior to the age five, if possible.

Asher and Olson (1983) reviewed 98 patients with spina bifida cystica, ranging in age from 5 years to 31 years of age, and looked at various factors which might affect their ambulation. They found that the most important variable affecting the ambulatory status of a patient with spina bifida cystica is the neurosegmental level of the paraplegia. Patients with third lumbar (influenced significantly by hip deformity) and higher levels of

paraplegia, usually became non-ambulatory, whereas patients with fourth lumbar or lower levels of paraplegia usually became functional walkers. Other variables such as age, obesity and some musculoskeletal deformities were found to be significant variables, and for this reason the prevention of obesity and musculoskeletal deformity is of importance.

Locally, Fraser et al.(1992) reviewed 55 patients with mid-lumbar myelomeningocele (L3 and L4) over a 17 year period, looking at factors which might affect hip stability and ability to walk. Two-thirds of the hips became dislocated or subluxed in the first year of life, more so in patients with an L3 level (86%) than with an L4 level (45%). The neurological level was also found to be the most significant determinant of walking ability, in that all patients with an L4 neurological lesion could walk, but only one third with an L3 neurological lesion could do so. Hip stability, IQ and fixed deformity did not influence walking ability.

Another factor, demonstrated by Samuelsson and Skoog (1988) from Stockholm, in a study of 163 patients, which may diminish walking ability is spasticity caused by syringohydromyelia or a Chiari malformation.

Various appliances have over the years been used to aid ambulation in patients with myelomeningocele. The use of the ankle foot orthosis in children with myelomeningocele with low lumbar and sacral lesions was evaluated by Thomas et al. (1989). Despite a small sample size they found that joint rotation was improved, with the ankle joint rotation being almost normal in 86% of the children and less so for rotation in the knee and hip joint.

Not all children who are ambulant remain so into adult life. Taylor and McNamara (1990) from Dublin reported on the ambulatory status of 87 adults with myelomeningocele who had been ambulant with calipers as children. Only 29 remained ambulant (23 community and 6 household ambulators). Of these 20 continued to use callipers, while 9 found they could walk well without them. All nonambulant

patients had discontinued wearing callipers as they found they were cumbersome and difficult to use and led to recurrent trouble with pressure sores.

The optimal method for providing mobility for children with high level myelomeningocele is controversial. Lough and Nielsen (1986) compared the use of the parapodium and a swivel walker in 10 patients with high level myelomeningocele. They found that walking velocities were significantly higher using the parapodium, but energy cost and gait efficiency were significantly better using the swivel walker. Liptak et al. (1992) compared children with high level myelomeningoceles, 39 of whom were using a parapodium with 29 using a wheelchair. They found the former group more likely to develop lesions of the lower extremities, to have dislocated hips, to be more obese and to watch more television, whereas the latter group were more likely to develop lesions of the gluteal region and to have knee-flexion contractures. The families found the parapodium only advantageous because it allows the upright posture. They suggested that a combined approach of using the upright posture and wheeled mobility to be optimal for these patients. Gajjar and Price (1988) reported on a locally designed vertical/prone wheeler which allows children with lower limb dysfunction the mobility of a wheelchair in the upright posture. Other than these advantages it is suggested that it can serve as an aid to strengthen and stabilise weight-bearing in children who are potential walkers with the aim of weaning the children onto a rollator with above-knee callipers and then hopefully crutches.

The incidence of scoliosis (a curve of >5% or more) in children with myelomeningocele is reported to vary between 9 and 88% depending on the age and the neurological level of the lesion (Shurtleff et al., 1974). Two main origins of scoliosis are recognised. The minority are congenital (usually associated with vertebral anomalies in addition to myelomeningocele), and the majority are developmental (due to inadequate neuromuscular control). Samuelsson and Eklof studied 163 patients with myelomeningocele of whom 143 had scoliosis (15 congenital). They found that the severity of the scoliosis increased

with age and was more severe the higher the level of the neurological deficit (the risk being considerably higher above a neurological level of L3).

The role of orthopaedic surgery in myelomeningocele appears to be specifically addressed at maintaining joint stability and achieving ambulation where possible. With time it has been realised that although it is possible to achieve ambulation in many children with myelomeningocele, there are those where it will be short-lived and requiring multiple surgical procedures (Asher and Olson, 1983; Taylor and McNamara, 1990). It appears therefore that ambulation is expected with a sensori-motor level below L2 and that hip instability plays an important role in ambulation especially in the mid-lumbar (L3 and L4) group, where the attention is directed (Hoffer et al., 1973; Asher and Olson, 1983; Fraser et al., 1992). Mobility in an upright position for infants with high neurological lesions can be achieved using a parapodium (Liptak et al., 1992) or a swivel walker (Lough and Nielsen, 1986) or a vertical wheeler which incorporates the advantages of the upright position and mobility (Gajjar and Price, 1988).

III.4.9 URINARY INCONTINENCE

The development of urinary tract disease in an infant born with myelomeningocele is well documented (Rose & Smith, 1963). Eckstein (1965) in his series of 148 children requiring a urinary diversion, 34 of them with myelomeningocele and a neurogenic bladder, found that 25% of the infants with myelomeningocele had gross dilatation of the upper urinary tract in the first two weeks of life. Smyth et al. (1974) found that 9 (9%) of 99 infants with lumbar or lumbosacral myelomeningocele had dilatation of the upper urinary tract and this was often associated with a neurogenic bladder. Jones and Williams (1967) in a series of 21 patients with myelomeningocele found two with hydronephrosis and 11 with bladder trabeculation or diverticula. Erikson et al. (1989) showed that urinary retention in the infant with myelomeningocele may be a transient consequence of the myelomeningocele repair and should be treated conservatively for at least 2 weeks

before any surgical intervention is considered. Nine of the 34 infants they studied who had normal renal sonograms preoperatively developed hydronephrosis and bladder distension postoperatively and all resolved on intermittent catheterisation over the next 2 weeks. Interest during the 1960's focused on urinary tract diversion both to preserve the upper urinary tract and to provide continence. This utilised an intestinal segment between the ureters and the skin, bypassing the bladder. Some authors recommended the procedure in every child over 2 years of age with a neurogenic bladder (Smith, 1966). The complication rate from this procedure, however, was noted with time to be unacceptably high and it was suggested that it be abandoned as a primary method of treatment (Light et al. 1977). The most common method of temporary urinary diversion in infants with a neurogenic bladder in myelomeningocele is a tubeless cutaneous vesicostomy. Allen (1980) reported the use of a vesicostomy as a temporary urinary diversion in 16 children less than 3 years of age with good results and felt that the operation was ideally suited for this purpose in this age group.

In 1966 sterile intermittent catheterisation was introduced as a way of managing the atonic bladder of hospitalised patients with spinal cord injuries (Guttmann & Frankel, 1966). Lapides et al. (1972) adapted the technique for other types of neurogenic bladders to a clean but nonsterile technique, which could in some cases be accomplished by the patient. This has become the basic method of care in most centres today. They studied 14 patients (12 females and 2 males) ranging in age from 3 to 65 years. An unspecified number had myelomeningocele. They showed an overall improvement using the technique. Kass et al. (1979) studied the use of clean intermittent catheterisation in children under 6 years of age. They studied 42 patients ranging in age from 1 week to 5 years of age, the majority with myelomeningocele. Twenty nine (69%) of the children were dry on this programme. Complications from the technique have been reported but are infrequent and include recurrent urinary tract infection, epididymitis and prostatitis (Kasabian et al., 1992), as well as urethral injury (Koleilat et al., 1989).

Kyker et al. (1977) compared intermittent catheterisation with a surgical diversion procedure. They reported on 52 patients with 26 in each group and found the preservation rate of the upper urinary tract to be high in both groups (92% vs 87%). Schoenberg and Meador (1982) followed 48 children on a clean intermittent catheterisation programme. They defined continence as being allowed less than two wet episodes per week. The children ranged from 3 to 15 years of age. In 25 (52%) good control was found and in a further 11 good control was possible if the children were more diligent. Of the 9 children who had dilated upper urinary tracts to start with, 6 had improved, 2 remained stable and only 1 deteriorated. Plunkett and Braren (1982) in their follow-up of 73 children, ranging in age from 11 months to 20 years, 47 of whom had myelomeningocele, found a low complication rate (7%) from the technique and recommended it as the treatment of choice in neurogenic vesical dysfunction where total bladder emptying is a problem.

More recently results of studies of intermittent catheterisation in children with myelomeningocele have shown variable results. Purcell and Gregory (1984) reported on 46 children being followed-up using clean intermittent urethral catheterisation. All patients were over 4 years of age and were given maximum therapeutic doses of supplemental medication to improve dryness. Fifteen (33%) were non-compliant, and only 11 (24%) were completely dry and wearing regular underclothes. Uehling et al. (1985) reported on 164 children, 85 of whom were on intermittent catheterisation and of these 53 had been on the programme for at least 5 years. They found 43 (81%) were dry, 49 (92%) had the occasional urinary tract infection and in only 7 (13%) had their renal status deteriorated on the programme. Their results are promising, but these patients were compliant and had been for at least 5 years. They do however, report on 21 failures to establish intermittent catheterisation as a satisfactory form of bladder management. Geraniotis et al. (1987) showed that the prophylactic use of clean intermittent catheterisation in 10 of 21 patients with bladder sphincter incoordination (the remainder were managed on self-voiding), ranging in age from 4 weeks to 6 months, prevented urinary tract deterioration (10% vs 50% in the self-voiding group).

Drug therapy may be required to enhance the effect of clean intermittent catheterisation. Mulcahy et al. (1977) reported on the use of oxybutynin chloride combined with intermittent clean catheterisation in the treatment of urinary incontinence in myelomeningocele patients. Of the 25 patients studied 21 (84%) achieved continence using this combination. Monitoring these drugs is important to avoid their frequent side effects, such as dry mouth, heat intolerance, sensitivity to strong light and mood change (Fernandes et al., 1991). Other medications such as calcium-channel blockers and potassium-channel agonist drugs are still under investigation (Hellstrom et al., 1989).

Bille et al. (1984) from Uppsala in Sweden showed that a bladder training programme consisting mainly of manual compression and or abdominal straining could achieve bladder emptying. They studied 43 children with myelomeningocele with complete urinary incontinence. One group commenced training before 3 years of age and the other group between 3 years and 13 years of age. Vesico-ureteric reflux was not considered a contraindication to the use of the method. Their results as regards continence were better starting the programme before 3 years of age. Of those with vesico-ureteric reflux 60% showed improvement on the method and it was suggested that patients not showing significant improvement should be candidates for clean intermittent catheterisation alone or with added drug therapy.

In some cases, where the urethral resistance is insufficient the placement of an artificial urinary sphincter may be necessary and in some centres is the procedure of choice (Gonzalez et al., 1989). Local experience regarding the placement of an artificial urinary sphincter is limited (Aaronson, 1986). Electrical bladder stimulation to increase bladder compliance and capacity has been used in very select patients in some centres (Decter et al., 1992).

It is accepted that continence in a child with myelomeningocele does not necessarily imply complete dryness all the time and that two wet episodes per week should be

accepted. The aim of urological management should be to prevent urinary tract damage and to achieve continence. Intermittent clean catheterisation is the simplest and least invasive way of achieving both these aims (Lapides et al., 1972). It has a low complication rate with a good success rate both for preservation of upper urinary tract and achieving continence (Plunkett and Braren, 1982), which is dependant on the compliance and diligence of the mother and/or child (Schoenberg and Meador., 1982). Drug therapy may be needed to enhance the effect of intermittent clean catheterisation (Mulcahy et al., 1977). There are failures on the programme and there are those neurogenic bladders more suitable to other modalities of treatment (cf. artificial sphincters; diversion procedures) (Gonzalez et al., 1989).

III.4.10 FAECAL INCONTINENCE

The majority of children born with myelomeningocele have limited control of their bowel actions. Forsythe and Kinley (1970) from Belfast treated 47 children with spina bifida who had faecal incontinence. The age of the children ranged from 2 to 10 years and their follow-up varied for 9 months to 3 years. They were managed on a regular toiletting programme on its own or in combination with enemas, suppositories and purgatives. They found that those children with normal anal tone were successful on the regular toiletting programme (17%), whilst others achieved continence using a combination of regular toiletting and enemas (32%), regular toiletting, enemas and suppositories (20%), and regular toiletting, enemas and purgatives (32%). Of the purgatives Senekot syrup proved to be the most economical and the most successful. Of the 15 patients on Senokot syrup all were continent, and they recommended it as an important method in the management of faecal incontinence in patients with myelomeningocele.

Dietrich and Okamoto (1982) studied 50 children with neurogenic bowel dysfunction and stool incontinence, 27 of whom had myelomeningocele, were all less than 19 years of age and of normal intelligence. All had received in-patient bowel regulation

training and were interviewed a year later. The frequency of stool incontinence had decreased from 1,8 episodes for the week prior to discharge to 0,9 episodes for the week prior to being interviewed. The use of bowel stimulants and high fibre diets had decreased, whereas dietary manipulation had increased and there had been significant changes in the recommended bowel programme in order to accommodate the individual home environment and family life styles. Of the children with myelomeningocele 15 (56%) had no incontinent episodes for the week preceding the interview.

Stellman et al. (1983) reported on the results of a questionnaire circulated to the parents of spina bifida children. A third (105) of all families contacted replied. Over 50% of these indicated that bowel management was a problem no matter what method of bowel management was used. Thirty five used manual evacuation, 22 were still in nappies, 22 were toiletting regularly, 12 were not using a formal programme, 11 used manual pressure and only 3 required the use of regular suppositories. Mothers bore most of the responsibilities regarding bowel management and 71% had no formal advice or training in bowel management. Families who complained of bowel management difficulties also had similar problems with bladder management.

Shandling and Gilmour (1987) from Toronto reported on the management of patients with spina bifida with faecal incontinence using a large-volume saline enema. To facilitate administration of the enema they devised a special catheter with a balloon to prevent leakage of the enema fluid. They reported on the use of the catheter in 112 patients over the age of 4 years all of whom had faecal incontinence. Excluding the four patients who abandoned the method, they claimed a 100% success rate and advocated its use for faecal incontinence in spina bifida.

Loening-Baucke et al.(1988) evaluated the efficacy of biofeedback training for faecal incontinence in patients with myelomeningocele. They randomised 12 patients to receive either conventional therapy alone, or in conjunction with biofeedback. Anorectal manometric functions were evaluated and 16 control children were also studied. They

found no difference between conventional therapy, where three of four patients reported a 75% improvement, and biofeedback, where three of eight patients reported a similar improvement.

In 1989 Willis from Victoria in Australia described a bowel washout programme similar to the one used in Toronto. He reported on 100 patients, 43 of whom had spina bifida. Thirty two of the spina bifida patients were still on the programme, 7 on daily washouts, 15 on two to three washouts per week, and 10 on weekly washouts to achieve reliable continence. Of the 11 who had stopped the programme 10 remained continent.

Blair et al. (1992) from Vancouver reported on the use of a balloon tipped catheter of their design. They studied 31 patients over a one year period, 19 of whom had myelomeningocele. They found the greatest improvement in children on the imperforate anus programme followed by those with myelomeningocele, where the mean number of incontinent episodes daily were significantly reduced from 3,1 before the use of their programme to 0,7 on their programme.

Liptak and Revell (1992) from New York evaluated 31 children, 30 of whom had myelomeningocele, using the Toronto enema continence catheter 18 to 20 months after commencing the programme. Fifteen patients dropped out of the study and of the remainder the proportion of continent stools rose significantly from 28% to 94% and of constipated stools dropped significantly from 55% to 15%. Although compliance was difficult for some families they suggested that it does provide significant improvement in the bowel care of selected patients with spinal cord impairment.

King et al. (1994) stressed the importance of education, patient compliance, age and the presence of anal reflexes in bowel training in spina bifida. They utilised a regular toiletting education programme on 40 children and young adults with spina bifida, which emphasised using diet, bulking agents, suppositories, digital stimulation, timed oral medication, and a consistent technique to induce bowel evacuation the same time each

day. Bowel continence, which was defined as one or fewer incontinent stools per month, rose from 13% to 60%. Compliance, the presence of bulbocavernosus and anocutaneous reflexes and instituting training before 7 years of age all improved the possibility of achieving continence.

Over the years the definition of faecal continence varies, which makes the comparison of studies difficult. The use of toiletting programmes to achieve continence do appear to achieve their goals in children with myelomeningocele (Forsythe and Kinley, 1970), but depend on the age of commencing the programme, the education of the child and parents in the programme, the compliance and the presence or absence of anal reflexes (King et al., 1994). The continence rate on a toiletting programme is not as high as has been reported by programmes using the bowel washout technique (Shandling and Gilman, 1987; Blair et al., 1992; Liptak and Revell, 1992). The role of biofeedback training in achieving faecal continence in patients with myelomeningocele is limited and needs future evaluation (Loening-Baucke et al., 1988).

III.5 CONCLUSIONS

Most first world countries have shown peaks and troughs, with a recent decline, for the prevalence of NTDs. The cause of the decline and whether this decline will continue is uncertain. The prevalence of NTDs in Africa and South Africa are poorly documented, and reports that have emanated are studies over short periods of time.

An increased prevalence of NTDs has been documented in conceptions that occur in winter or early spring, in the offspring of women at the extremes of their reproductive life, in female infants, in first borns and amongst higher birth order, and those from a low socio-economic class, by some investigators but not others.

The role of the fungus Phytophora infestans has been incriminated as an aetiological agent in the past but never proven. The protective effect of supplemental folic acid in preventing NTDs and their recurrence has been shown and it has been suggested that folic acid metabolism may differ in women at risk of having a NTD affected pregnancy. Following these studies it has been recommended that all women of child bearing age who are capable of becoming pregnant should receive folic acid supplementation.

The diagnosis of neural tube defects, in particular anencephaly and open myelomeningocele, can be made prior to twenty weeks gestation antenatally, using maternal serum alpha-fetoprotein and/or acetylcholinesterase, afterwhich the reliability of the test diminishes as the natural surge in these products occurs. Antenatal sonography, has been refined over the years and a detailed fetal sonograph is now used in preference to the alpha-fetoprotein screen at 18 weeks gestation in some centres. The success of a screening programme depends on the compliance of the attending public and the avilability of resources, the costs involved in running the programme and the equipment involved. Locally, most mothers attend antenatal for the first time well into their second trimester making screening difficult. Although sonographic facilities are widely available in Cape Town and South Africa, high resolution fetal sonography has only recently been available and at a few large centres.

The role played by the mode of delivery in the outcome of a myelomeningocele pregnancy is uncertain. The role of caesarean section is as yet unsettled in myelomeningocele, except in a breech presentation and where other obstetric indications exist.

Survival rates have improved with improved neurosurgical techniques and advances in the control of hydrocephalus. Most studies agree that early closure (<48 hours) improves survival and prognosis and reduces the number of infection related deaths. The argument as to whether early closure improves lower limb muscle power as suggested by Sharrard et al. (1963) has not been substantiated by others.

Since the introduction of selection criteria, the results of unselected series have shown an improvement in survival and outcome as the surgical, urological and medical care of these patients has improved to give results, which when the results of some studies are considered, are very similar to the outcome of selected series. Questions regarding selection criteria have arisen. Reports that the motor function in lower limbs could improve up to 10 days post delivery, with or without surgery, have brought the criterion of paraplegia at birth into question, and that poor intellectual development was linked to CNS infection and not necessarily only hydrocephalus, questions the role of hydrocephalus as a selection criterion. The multiple handicaps seen in myelomeningocele have been well documented and the role played by the extent of the neurological deficit and IQ in determining disability and dependency emphasised.

Selection has shown an improved outcome for children with myelomeningocele, but at a price. Including those selected for no surgical intervention, the mortality rates are high (60%). However, their intellectual outcome for normal IQ's is approximately 15% better than for unselected series. Whether this marginal increase in intelligence is worth the increase in mortality is questionable. Families, who are frequently asked to help in the decision making process, and the nursing staff who have to nurse the untreated patients through difficult days until their demise, are also affected by selection. With advances and improved outcome reported from more recent unselected series, it is difficult to justify selection purely on the grounds of an improved outcome.

Predicting outcome in the case of myelomeningocele is not an easy task. Although intellectual outcome appears to be the area which influenced predictions more than future limb paralysis, it proved to be difficult to predict, especially when associated with hydrocephalus.

Children selected out for conservative care only, may well live far longer in their own home environment and allow the parents time to bond, care and eventually mourn for their baby more successfully. The role of the family's socio-economic status in the survival

and outcome of infants with myelomeningocele has been questioned. Very few studies have looked at cross-cultural customs, traditions and feelings with regard to a baby with a myelomeningocele. In Africa, the way western medicine may be in conflict with these beliefs and views is important to understand.

Ventriculo-peritoneal shunts are the preferred procedure in the management of hydrocephalus in myelomeningocele, but simultaneous shunt placement with lesion closure which would save the child a second operation and anaesthetic has been suggested by some. Shunt failure appears to be greater in those shunts placed in the first year of life and surveillance should be high during this period. While the anterior fontanelle remains patent, cranial sonography is now considered an easy non-invasive method of diagnosing and monitoring hydrocephalus.

The presence of hydrocephalus and CNS infection appear to be the two major determinants of intellectual outcome in myelomeningocele, the latter being the more important. The absence of hydrocephalus in myelomeningocele allows for a better prognosis, but compared to their siblings, although functioning in the normal range for intelligence, they do not do as well. Some earlier studies were optimistic about intellectual outcome, especially those using variable follow-up periods, using different assessments in the various age groups which may have led to the optimism.

Orthopaedic surgery in myelomeningocele is addressed at maintaining joint stability and achieving ambulation where possible. Although it is possible to achieve ambulation in many children with myelomeningocele, in some it will be short-lived, requiring multiple surgical procedures. Ambulation is expected with a sensori-motor level below L2 and hip instability plays an important role especially in the mid-lumbar (L3 and L4) group, where the attention is directed. Mobility in an upright position for those with high neurological lesions can be achieved using a parapodium or a swivel walker or a vertical wheeler which incorporates the advantages of the upright position and mobility.

Continence in a child with myelomeningocele does not necessarily imply complete dryness at all the time and that two wet episodes per week should be accepted. Other than to achieve continence, the aim of urological management should be to prevent urinary tract damage. Intermittent clean catheterisation is the simplest and least invasive way of achieving both these aims. Complication rate is low with a good success rate both for preservation of upper urinary tract and achieving continence, which depends on the compliance and diligence of the mother and/or child. Drug therapy may be needed to enhance the effect of intermittent clean catheterisation. There are failures on the programme and there are those neurogenic bladders more suitable to other modalities of treatment (cf. artificial sphincters; diversion procedures).

Over the years the definition of faecal continence varies, which makes the comparison of studies difficult. The use of toiletting programmes to achieve continence do appear to achieve their goals in children with myelomeningocele, but depend on the age of commencing the programme, the education of the child and parents in the programme, the compliance and the presence or absence of anal reflexes. The continence rate is not as high as has been reported by programmes using the bowel washout technique. The role of biofeedback training in achieving faecal continence in patients with myelomeningocele is limited and needs further evaluation.

IV METHODOLOGY

IV.1 PREVALENCE

IV.1.1 PREVALENCE AND SECULAR TRENDS

All cases of anencephaly, inencephaly, encephalocele and spina bifida in infants delivered in the greater Cape Town area, during the period 1st January 1973 to 31st December 1992 were sought. The category 'spina bifida' included all defects from myelocele to meningocele. Greater Cape Town for the purposes of this study comprised the Cape Town Municipality (population 942 997; 1992) and the rapidly developing periurban settlement of Khayelitsha. This settlement started in 1983 and by 1990 was officially estimated to have a population of 184 235. This figure was felt to be an underestimate and general consensus placed the population between 300 000 and 350 000 (Harrison and McQueen, 1992).

The birth data for the Cape Town Municipality were obtained from the office of the Medical Officer of Health (MOH). The Peninsula Maternal and Neonatal Service (PMNS) of the Department of Obstetrics and Gynaecology of the University of Cape Town serves the lower socio-economic sector of the population of the city. The more affluent section of the population delivers privately, but all births within the metropolitan area are reported to the MOH. Mothers living in Khayelitsha deliver within the PMNS. In 1991/2 69% delivered in a Midwife Obstetric Unit (MOU), 27% were referred to a maternity hospital and 5% were born at home. The denominator data for the study were a combination of the MOH birth records plus the PMNS births for Khayelitsha.

The records reviewed included:

(i) The following maternity hospitals:

- Groote Schuur Hospital (Maternity Centre);

- Mowbray Maternity Hospital;
- New Somerset Hospital;
- Peninsula Maternity Hospital;
- St. Monica's Maternity Hospital;

(ii) The following Neurosurgical units:

- Groote Schuur Hospital;
- The Red Cross War Memorial Children's Hospital;

(iii) The Fetal Abnormality Group based at the Maternity Centre at Groote Schuur Hospital, which started in 1983. This consists of a group of perinatalogists (Obstetricians, Neonatologists, Ultrasonologists and Geneticists) who review all fetal abnormalities diagnosed antenatally:

- from within the Peninsula Maternal and Neonatal Service;
- referred from private practice for confirmation only and or further management;
- referred from other centres outside the Cape Town area;
- referred for termination of pregnancy for fetal reasons.

(iv) The Department of Human Genetics at the University of Cape Town;

(v) The Spinal Defect's Clinic at the Red Cross War Memorial Children's Hospital;

(vi) The Regional Offices for the registration of deaths of the Department of Home Affairs in Cape Town and Observatory.

The number of infants delivered with a neural tube defect per calendar year and the prevalence of the condition were calculated per calendar year.

IV.1.2 MATERNAL AGE

The maternal age of all cases of a neural tube defect were recorded. The number of infants delivered with a neural tube defect per year of maternal age was determined and the prevalence of the condition calculated per year and per five year period of maternal age.

IV.1.3 SEASONAL PREVALENCE

The date of conception was calculated for all infants. For infants delivered preterm this was calculated from the scored gestational age (Ballard et al., 1979) or from antenatal sonographic findings. Where this evidence was unavailable the estimated gestational age recorded in the birth chart was used. In the case of still birth or termination of pregnancy the gestational age was calculated from antenatal ultrasonic findings, if available, or if not the estimated gestational age recorded in the birth chart was used. The prevalence of neural tube defects per month and per season of conception was then calculated.

IV.1.4 SEX RATIO

The sex of all affected cases, where possible, was recorded. This was expressed as a annual and overall ratio of male to female for individual and total neural tube defects for the period.

IV.1.5 PARITY

The parity of all mothers was recorded, and the prevalence of neural tube defects in birth order was determined.

IV.1.6 STATISTICS

Using the LR (Logistic Regression) program of the BMDP (Biomedical Data Package), a stepwise logistic regression model was applied to the yearly data of prevalence of anencephaly, encephalocele, myelomeningocele and other forms of spina bifida, differentiated on the basis of race (White, Mixed Ancestry, Black) (editor Dixon, 1988). Non parametric methods using 3S in BMDP were applied to two-way classifications of prevalence rates, to seek evidence of any consistency of prevalence across neural tube defects, races and seasons of conception.

The choice of logistic instead of Poisson regression was because it can be shown that for events with small probabilities of occurrence, application of logistic regression to the event frequencies relative to the known total of the sub-population, yields the same estimates for regression coefficients and their standard deviation errors, as application of Poisson regression (SAS Manual, 1989).

IV.2 OUTCOME

IV.2.1 PATIENTS

All patients who underwent surgery for a congenital myelomeningocoele from 1 January 1979 to 31 December 1985, within the Department of Neurosurgery of the University of Cape Town, either at The Red Cross War Memorial Children's Hospital or at Groote Schuur Hospital, qualified for admission into the study.

IV.2.2 ANTENATAL DATA (Appendix 2)

All mothers attending the Spinal Defect's Clinic at the Red Cross War Memorial Children's Hospital were interviewed. The following details were obtained from the mother:

- Maternal Age, at the time of affected pregnancy;
- Social Class (according to Molteno et al., 1980)
- Maternal Education, highest standard reached;
- Parity, at time of affected pregnancy;
- Gravidity, at time of affected pregnancy;
- Family History of Neural Tube Defects;
- Antenatal Alpha-Fetoprotein Studies;
- Antenatal Sonography Studies;
- Antenatal Diagnosis, during affected pregnancy;
- Place of Delivery.

Social class was categorized according to the occupation of the breadwinner. Molteno et al. (1980) found that for the local population of mixed ancestry a three division social class was appropriate (social class I - III - 36%; IV - 27%; V - 36%). As the majority of our patients are of mixed ancestry the same classification was used. Social class I

represents the United Kingdom General Classification of social class I - III, social class II social class IV and social class III social class V (Registrar General, 1960).

IV.2.3 NEONATAL DATA (Appendix 2)

The following information was obtained from the birth records or from the 'Road to Health Card' if the former was unavailable:

- Mode of Delivery;
- Birth Weight;
- Head Circumference;
- Gestational Age;
- Apgar Scores;
- Risk Factors.

All infants were clinically examined by a member of the neurosurgical medical staff and the following information was obtained from the patient records:

- Sex of Infant;
- Site of Lesion;
- Size of Lesion;
- Evidence of Sepsis;
- Presence of Other Congenital Malformations;
- Sensori-Motor Level;
- Presence of Fixed Bony Abnormalities of the Spine or Legs;
- Hip Location;
- Anal tone;
- Timing of Surgery;
- Nature of Surgery Undertaken; &
- The Presence or Absence of any Obstructive Uropathy.

IV.3 THE FOLLOW UP STUDY

IV.3.1 CLINICAL ASSESSMENT

The children were seen on an annual basis at the Spinal Defect's Clinic at the Red Cross War Memorial Children's Hospital. The children were clinically examined and any neurosurgical, orthopaedic, urological and surgical events were recorded. Sensori-motor levels in the lower limbs, methods of ambulation and attaining urinary and faecal continence were monitored.

IV.3.2 DEVELOPMENTAL ASSESSMENT

The children were developmentally screened using a rapid developmental screening test (DEI) devised and used by the Developmental and Assessment Clinic at the Red Cross War Memorial Children's Hospital. The clinical examination and these assessments were done by either the author or the former head of the Developmental and Assessment Clinic (Prof. C.D. Molteno).

IV.3.3 THE GRIFFITHS ASSESSMENT

The test administered was the standardised Griffiths Mental Developmental Extended Scales (0 - 8 years), using a set of standardised test equipment, purchased from The Test Agency in London (Griffiths, 1970). The scales are widely used in the United Kingdom and South Africa (Hanson and Alridge Smith, 1982). They have been found to correlate well with the Bayley Scales (Ramsay and Fitzhardinge, 1980), the Stanford Binet (Ludlow and Allen, 1979) and the Cattell Infant Intellectual Scales (Caldwell and Drachman, 1964). The scales have been standardised for local conditions (Allan et al.,1989, Luiz, 1988), and the Griffiths developmental quotients correlate highly with the

Junior South African Intelligence Scales (JSAIS) (Heimes 1983). They have been used to evaluate handicapped children, both Down Syndrome (Ramsey and Fitzhardinge, 1980; Ludlow and Allen, 1979; and Neser et al., 1989) and spina bifida (Welbourn, 1975). Welbourn (1975) used the Stanford-Binet and the Griffiths to select spina bifida children for normal schools or special facilities, but preferred the Griffiths Scales because they were compact and convenient to administer and report on.

The Scales were administered by the author, who is a registered user of the Griffiths Mental and Developmental Extended Scales (0 - 8 years). The test results were recorded in the standardised Griffiths record book (blue book). This was performed within one calender month of the child's fifth birthday. The test was performed in the mornings and in an appropriate environment, with no distractions, at the Developmental and Assessment Clinic (away from the clinical milieu associated with surgery or follow up procedures). The test was conducted in the child's home language.

The Griffiths Mental and Developmental Extended Scales (0 - 8 years), consists of six subscales, namely, the locomotor scale, the personal-social scale, the hearing and speech scale, the hand and eye scale, the performance scale and the practical reasoning scale.

IV.3.3.1 THE LOCOMOTOR SUBSCALE

This subscale gives opportunity to observe certain physical weaknesses or disabilities, or more definite defects of movement in young children. Items include the ability to run fast out of doors, to bounce and catch a ball, to jump over a 15-25 cm rope etc.

IV.3.3.2 THE PERSONAL-SOCIAL SUBSCALE

This subscale gives opportunity to assess personal and social development. Items include the ability to give his/her home address, to dress and undress self, to fasten buckles, tie a knot etc.

IV.3.3.3 THE HEARING AND SPEECH SUBSCALE

This subscale is the most intellectual of the scales and gives opportunity for the study of the growth and development of language. Items include naming colours, comprehension of items, opposites, repitition of sentences with 6-16 syllables etc.

IV.3.3.4 THE EYE AND HAND CO-ORDINATION SUBSCALE

This subscale consists of items relating to the handwork and visual ability of the child. Items include drawing, writing, threading beads, etc.

IV.3.3.5 THE PERFORMANCE SUBSCALE

This subscale is very largely a scale of performance tests and enables the examiner to observe and measure skill in manipulation, speed of working and precision. Items include formboards, pattern making etc.

IV.3.3.6 THE PRACTICAL REASONING SUBSCALE

This subscale concentrates mainly on recording the earliest indications of arithmetical comprehension, and the realisation of the simplest practical problems. It indicates the child's ability to benefit from formal schooling.

IV.3.4 SCORING THE EXTENDED SCALES

In this age category each item within each subscale carries two months of mental age (M.A.) credit. A profile is finally obtained by dividing each of the subscale M.A. credits by the chronological age (C.A.) and multplying by 100, to obtain the separate sub-quotients. The final General Quotient (G.Q.) is the average of these six separately obtained quotients. As a number of children with high lesions would obtain a lower G.Q. because of their locomotor paralysis, and this would not be a fair reflection of their true intellectual capabilities when compared to the children with lower lesions, a Modified General Quotient (M.G.Q.), excluding the Locomotor Subscale, was calculated for all the children studied (Heimes 1983).

IV.3.5 METHODS OF DATA RECORDING AND ANALYSIS

IV.3.5.1 DATA RECORDING

The hospital number, all antenatal and neonatal data were entered onto an information sheet. In addition, when the follow up data were also collected, this was added to the information sheet. Finally, when the Griffiths data were collected, they too were entered onto the information sheet. These data were later computerised for further analysis. The data were then reviewed and checked for transcription errors.

IV.3.5.2 DATA ANALYSIS

The coded data were analysed by computor. The analyses were performed using the Epi. Info. (version 5) - a Word Processing Data Base and Statistics System for Epidemiology on Microcomputors, from the U.S. Department of Health and Human Services at the Centre for Disease Control, Atlanta, Georgia.

Further data analysis data was performed using the LR (Logistic Regression) program of the BMDP (Biomedical Data Package)(ed. Dixon, '88), and a stepwise logistic regression model was applied. The binary response variables ambulatory status, bladder management, and modified Griffiths Quotients were each separately examined and evidence was sought of any useful explanatory variables amongst those recorded, and of any important relationships arising from the interaction of explanatory variables. The size of the data set (n=80) necessarily limits the complexity of the interactive models that can be applied.

V RESULTS

V.1 PREVALENCE

During this period 516 252 deliveries took place in the Cape Town area (white, 10%; mixed ancestry, 60%; black, 30%) of which 606 had neural tube defects (NTD). Of these deliveries 401 had spina bifida. Three hundred and thirty nine had myelomeningocele, 34 (10%) of which were legally terminated. There were 25 (7%) stillbirths, 87 (26%) were neonatal deaths and 17 (5%) were deaths subsequent to treatment. One hundred and seventy six (52%) survived infancy (Table V.1.1.).

TABLE V.1.1 NEURAL TUBE DEFECTS (NTD) BY RACE IN CAPE TOWN, 1973 - 1992

	RACE			TOTAL(%)
	W	MA	B	
NEURAL TUBE DEFECTS	133	325	148	606(100)
ANENCEPHALY	36	95	33	164(27)*
ENCEPHALOCELE	11	21	9	41(7)
SPINA BIDIFA	86	209	106	401(66)
MYELOMENINGOCELE	73	172	94	339(56)
OTHER	13	37	12	62(10)

* - includes 9 cases of iniencephaly (2W; 4 MA; 3B)

W - White; MA - Mixed Ancestry; B - Black

V.1.1 PREVALENCE AND SECULAR TRENDS

During this period the prevalence of neural tube defects fluctuated but no statistical trend could be demonstrated. Times of increased frequency were noted in 1975 (1,53/1 000 births), 1977 (1,72/1 000 births) and 1985 (1,74/1 000 births), and in 1990 (1,40/1 000 births). Years of low prevalence were recorded in 1986 (0,80/1 000 births), 1987 (0,79/1 000 births) and in 1992 (0,63/1 000 births) (Figure V.1.1).

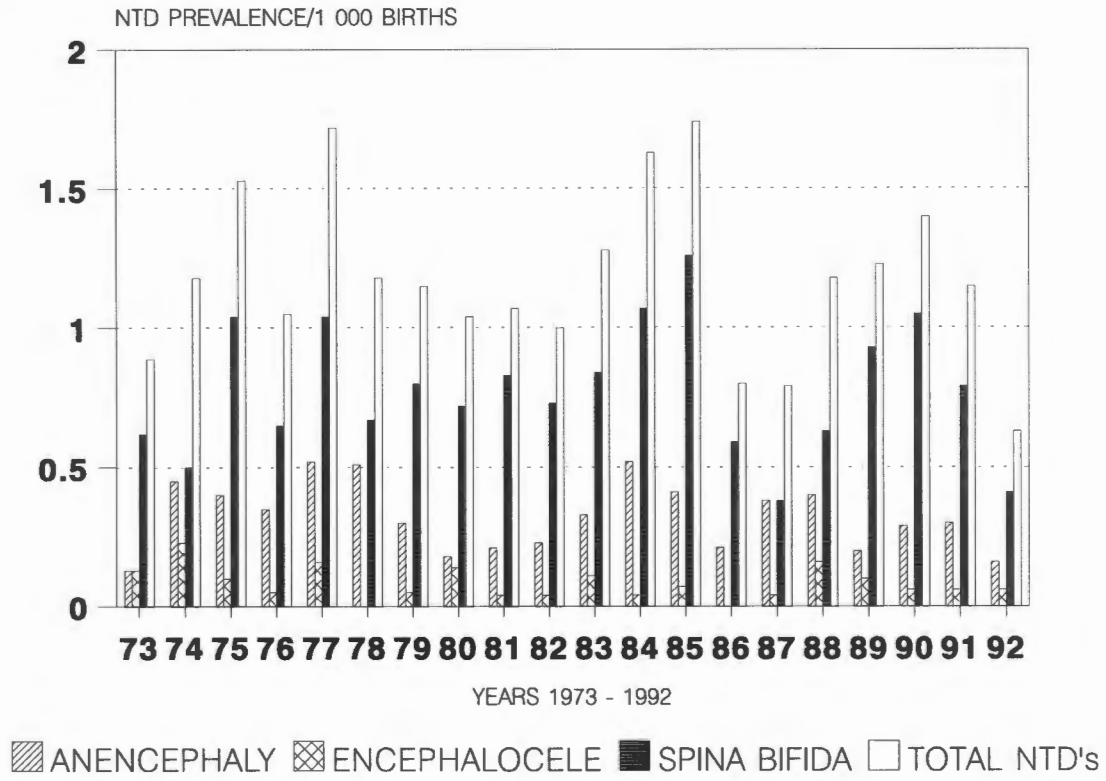

Fig.V.1.1. The prevalence of neural tube defects (NTD) in Cape Town, 1973 - 1992.

These patterns were demonstrated for spina bifida but not so for anencephaly. A Friedman two-way analysis of variance of ranks gave no statistical evidence for common time patterns across neural tube defect groups.

The overall prevalence of all forms of NTDs for the 20 year period was highest amongst whites and lowest amongst blacks with the exception of myelomeningocele where the lowest prevalence was amongst those of mixed ancestry.

Neural tube defect prevalence fluctuated over the period more so in the white population group than for the other groups (Figure V.1.2).

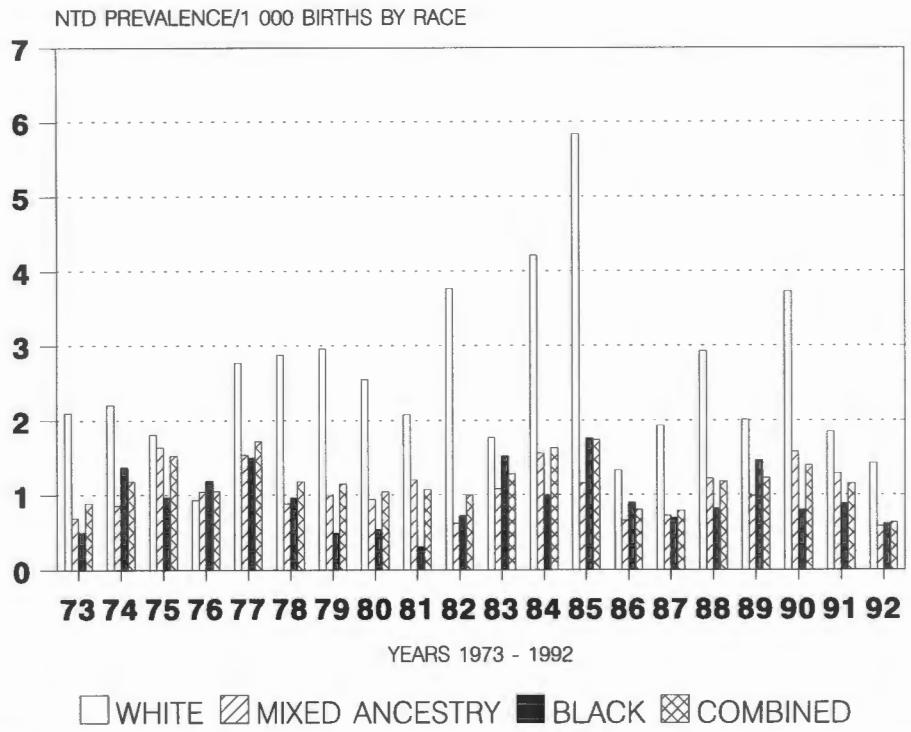

Fig V.1.2 The prevalence of neural tube defects (NTD) by race in Cape Town, 1973 - 1992.

In 1985 the rate in whites reached a peak of 5,84/1 000 births. The overall prevalences for the 20 year period for the various racial groups in Cape Town are :

Whites - 2,56/1 000 births;

Blacks - 0,95/1 000 births;

Mixed Ancestry - 1,05/1 000 births.

Similarly, Friedman two-way analysis of variance gave no evidence of common time patterns across race groups. The ratio of spina bifida to anencephaly was 2,39 for whites, 3,21 for blacks, 2,2 for persons of mixed ancestry, and 2,45 for the combined group.

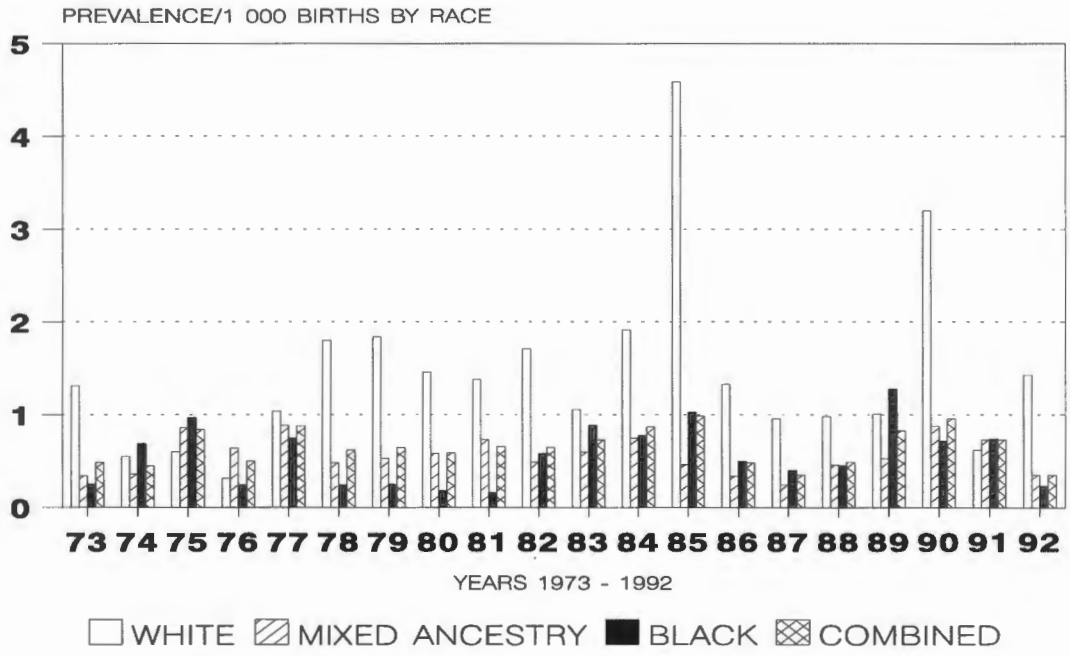

Fig.V.1.3. The prevalence of myelomeningocele by race in Cape Town, 1973 - 1992.

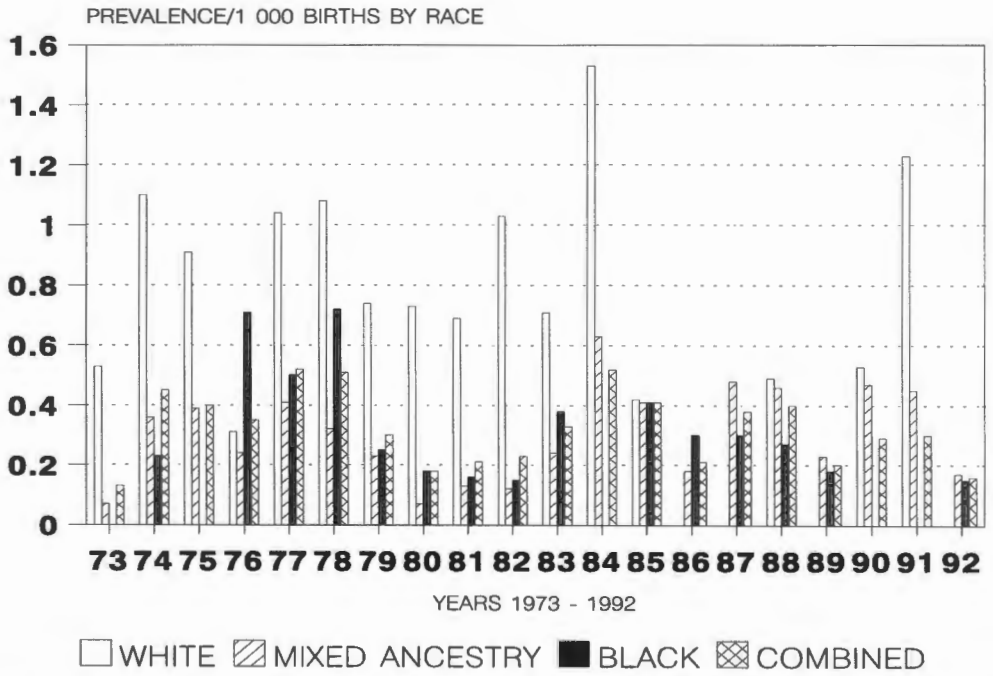

Fig.V.1.4. The prevalence of anencephaly by race in Cape Town, 1973 - 1992.

the prevalence has fluctuated considerably over the 20 year period for all race groups but more so for the white group for myelomeningocele.

The models that were examined using the BMDP LR program sought evidence for race differences and changes over time. There were no statistically significant linear changes in time in the log odds terms. This finding implies that the annual data may be viewed as similar multiple observations on the same phenomenon. However, the analysis exhibited significant effects on the log odds ratios associated with race, in that prevalence was significantly higher amongst the white group. Generally, the odds ratio (p/1-p) was two to three times larger than the corresponding figures for the mixed ancestry and black groups

(Table V.1.2).

TABLE V.1.2 ODDS RATIOS (OR) AND CONFIDENCE INTERVALS (CI) FOR ODDS RATIOS FOR FREQUENCY OF NEURAL TUBE DEFECTS (NTD) BETWEEN RACE GROUPS IN CAPE TOWN, 1973 - 1992

	p FOR OBSERVED F RATIO RACE DIFFERENCES	WHITE vs MIXED ANC		WHITE vs BLACK	
		OR +	CI	OR +	CI
ANENCEPHALY	<0,0001	2,26	1,52 - 3,27	3,38	2,10 - 5,44
MYELOMENINGOCELE	<0,0000	2,46	1,87 - 3,24	2,32	1,71 - 3, 17
OTHER	<0,0002	2,35	1,34 - 4,12	3,93	1,91 - 8,09
SPINA BIFIDA	<0,0000	2,44	1,90 - 3,12	2,52	1,90 - 3,35
TOTAL NTD's	<0,0000	2,37	1,93 - 2,92	2,73	2,14 - 3,47

V.1.2 MATERNAL AGE

The number of individuals with neural tube defects decreased with increasing maternal age, except for a peak in the early twenties, but the prevalence appears to peak at both extremes of the maternal age span. If the data are presented in five year periods, the peak effect appears more clear cut (Figure V.1.5).

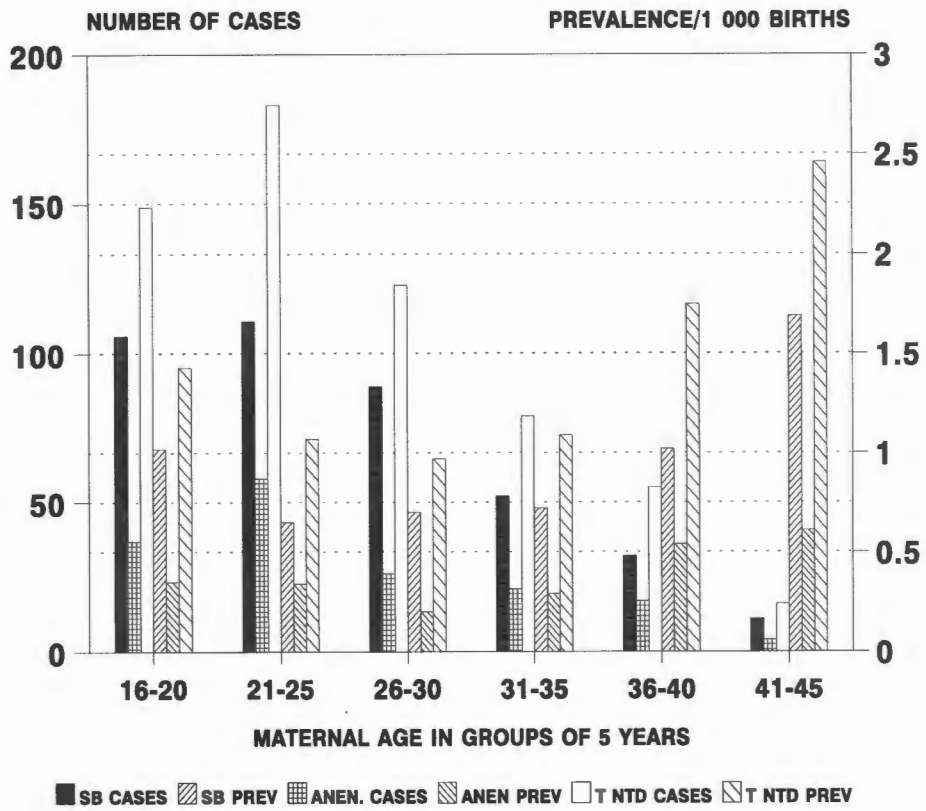

Fig.V.1.5. Prevalence of Neural tube defects vs maternal age group in Cape Town 1973 - 1992

However, the prevalence for the 41-45 year age group is subject to greater variation and the observed effect may be an artefact of data collection. The pattern of maternal age distribution does not exhibit systematic variation, but differences between race groups did exist and hence adjusted figures for race and not time are presented, where applicable.

V.1.3 SEASONAL VARIATIONS

A peak in the conception of cases of neural tube defects occurred during the months from July to September. This finding is similar for whites and blacks, whereas the profile is somewhat flatter for cases of mixed ancestry. A Friedman test indicates that the consistency of the observed pattern over months is statistically significant. When the data are

assessed in seasons the possible cyclical effect is more obvious (Figure V.1.6).

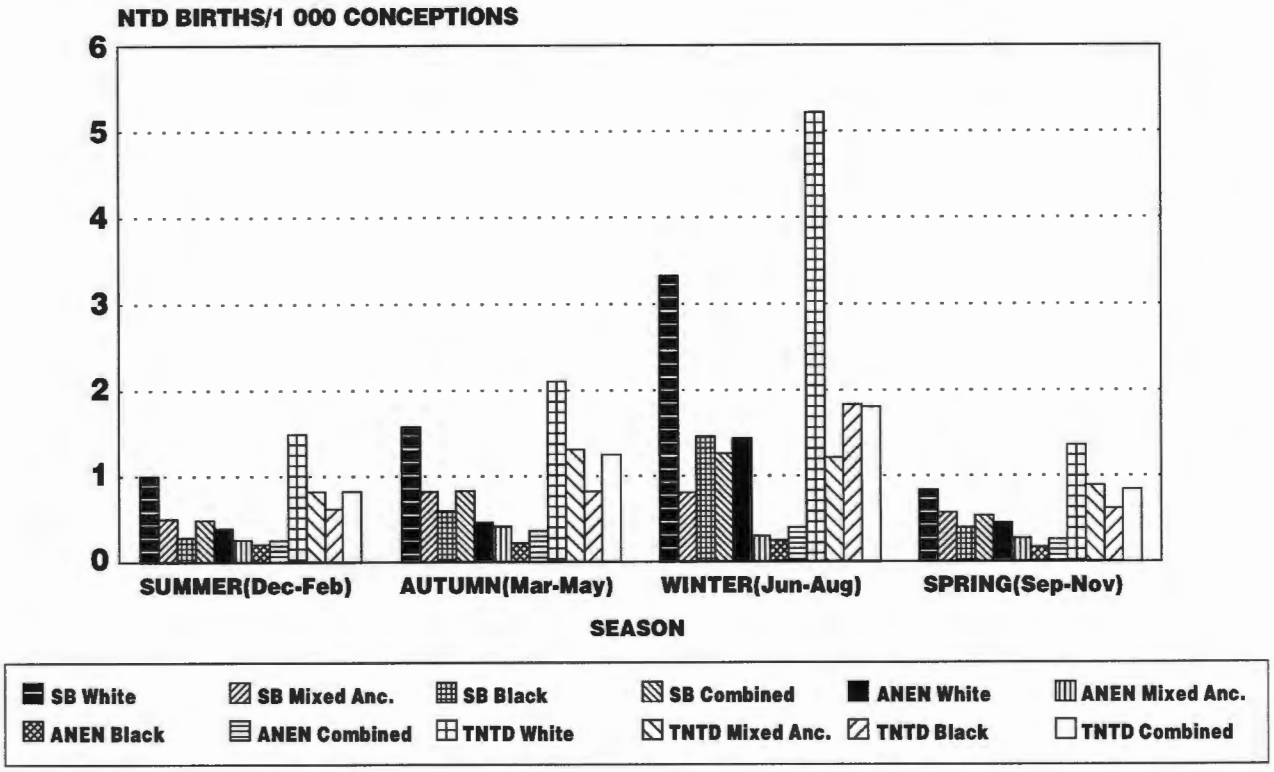

Fig. V.1.6. Prevalence of NTD births by season of conception in Cape Town, 1973 - 1992 (SB - Spina Bifida; Anen - Anencephaly; TNTD - Total NTD; Anc. - Ancestry)

V.1.4 SEX RATIO

The sex of NTD cases was indeterminate for three infants because of extreme prematurity and unrecorded in a further five fetuses at termination of pregnancy. Overall, there is a female predominance for all neural tube defects. The male to female ratios are 0,89 for spina bifida, 0,93 for myelomeningocele, 0,68 for other forms of spina bifida, 0,67 for anencephaly and 0,82 for total NTD. On a racial basis, the white and black groups show a slight female predominance for spina bifida, myelomeningocele, and other forms of spina bifida, which overall has shown a variation in the sex ratio over the period of study (Table V.1.3).

TABLE V.1.3 SEX RATIOS (M:F) BY RACE FOR NEURAL TUBE DEFECTS (NTD) IN CAPE TOWN, 1973 - 1992

	WHITE	BLACK	MIXED ANCESTRY	OVERALL
SPINA BIFIDA	1,03	1,08	0,74	0,89
MYELOMENINGOCELE	1,06	1,09	0,81	
OTHER	1,17	1,00	0,48	
ANENCEPHALY	0,78	0,53	0,68	0,67*
ENCEPHALOCELE	0,57	0,80	0,75	0,71
TOTAL NTD's	0,97	0,95	0,72*	0,82*

* - significantly different from 1 at 5% level

V.1.5 PARITY

The prevalence of spina bifida, anencephaly and hence total NTD are highest at the extremes of birth order in affected families (Figure V.1.7).

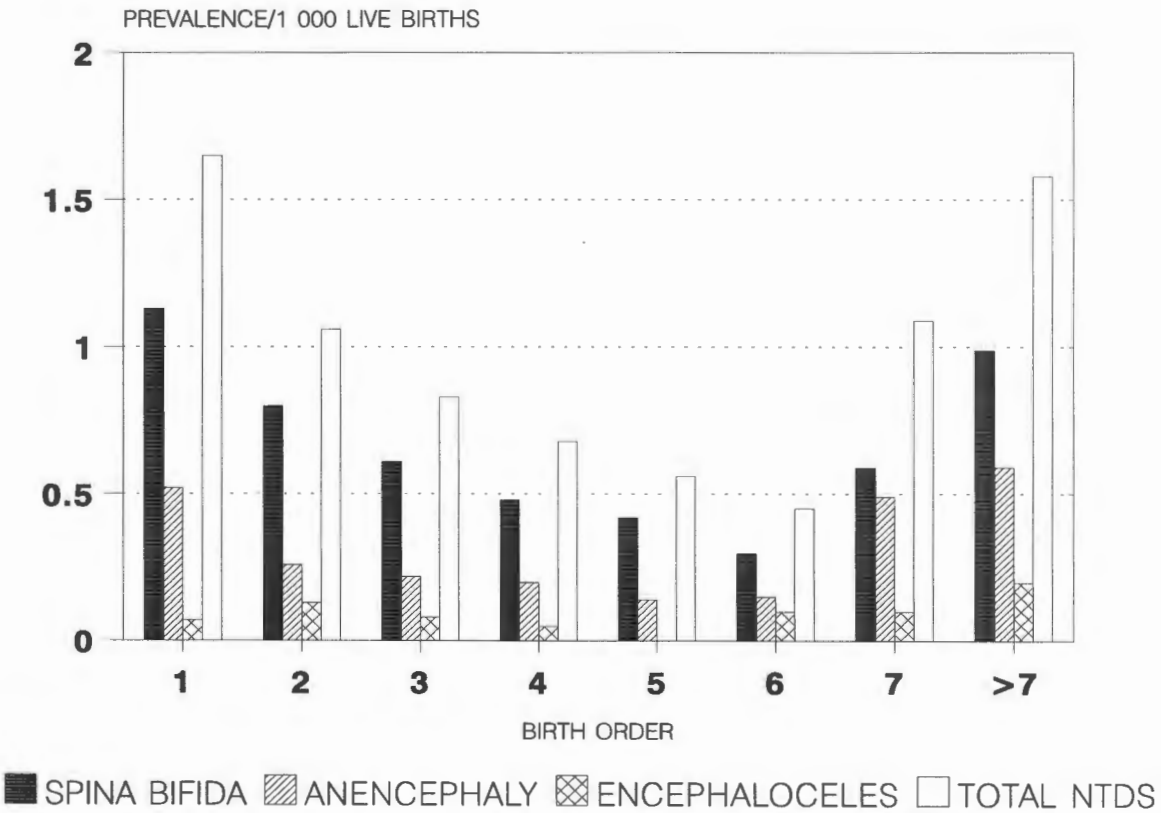

Fig V.1.7. The prevalence of neural tube defects (NTD) versus birth order in Cape Town, 1973 - 1992.

There is a decline during the mid-range of birth order. The prevalence for anencephaly is much lower than that for spina bifida, but a similar trend may be present. This trend is not shown for encephaloceles. A Friedman test for inconsistencies between spina bifida and anencephaly is not significant, that is the observed trends are consistent.

V.2 OUTCOME

During the study period 118 infants underwent surgery for a congenital myelomeningocele. The majority were of mixed ancestry (25 White; 64 of Mixed Ancestry; 29 Black), and there was a male predominance (64 males; 54 females). Similar numbers of infants from the White and Mixed Ancestry groups came from both the greater Cape Town area and from outside of Cape Town, but significantly more infants from the Black group came from outside of Cape Town (Table V.2.1).

Table V.2.1 RACE AND SEX DISTRIBUTION

	BIRTH ORIGIN		
RACE/SEX GROUP	GREATER CAPE TOWN	OUTSIDE CAPE TOWN	TOTAL
	n (%)	n (%)	n (%)
Black	7 (13,2)	22 (33,9)	29 (24,6) #
M/F	*6/1*	*7/15*	*13/16*
Mixed Ancestry	32 (60,4)	32 (49,2)	64 (54,2) #
M/F	*18/14*	*18/14*	*36/28*
White	14 (26,4)	11 (16,9)	25 (21,2) #
M/F	*8/6*	*7/4*	*15/10*
TOTAL	53 (100)	65 (100)	118 (100)
M/F	*32/21*	*32/33*	*64/54*

M/F - Male / Female; # - $p < 0,05$

The majority of the mothers came from Social Class III (49,2%), however, approximately a third came from Social Class I (36,4%). There was a social class difference between the group from the greater Cape Town area and those from the outside of Cape Town, in that more mothers were of Social Class I (27 vs 16) and fewer of the Social Class III (18 vs 40) in the greater Cape Town group (Table V.2.2).

Table V.2.2 SOCIAL CLASS DISTRIBUTION

SOCIAL CLASS	BIRTH ORIGIN		TOTAL
	GREATER CAPE TOWN	OUTSIDE CAPE TOWN	
	n (%)	n (%)	n (%)
I	27 (50,9)	16 (24,6)	43 (36,4)#
II	8 (15.1)	9 (13,9)	17 (14,4)#
III	18 (34,0)	40 (61,5)	58 (49,2)#
TOTAL	53 (100)	65 (100)	118 (100)

- $p < 0,01$

The majority of the mothers were in their twenties (57,6%) and thirties (24,6%). The maternal age distribution in the greater Cape Town area was similar to that of patients coming from outside of Cape Town. There were five mothers who were over forty years of age, the oldest being 55 years of age, and of rural origin. Of the sixteen teenage pregnancies more came from the Cape Town area and only one came from a social class I background (Table V.2.3).

The educational profile for the mothers from the greater Cape Town area differed from those who came from outside of the Cape Town area, in that the mother from the greater Cape Town area had a better standard of education, with more attending secondary school (17 vs 12), matriculating (15 vs 9) and receiving tertiary education (3 vs 1). Thirty (25,4%) of the mothers had no formal education, 24 (80%) of whom came from outside of the greater Cape Town area (Table V.2.4).

Table V.2.3 MATERNAL AGE VS SOCIAL CLASS

MATERNAL AGE	SOCIAL CLASS I	II	III	TOTAL
	n (%)	n (%)	n (%)	n (%)
< 19 years	1 (2,3)	2 (11,8)	13 (22,4)	16 (13,6)#
CT/NON-CT	*1/0*	*1/1*	*8/5*	*10/6#*
20-29 years	33 (76,7)	10 (58,8)	25 (43,1)	68 (57,6)#
CT/NON-CT	*19/14*	*6/4*	*5/20*	*30/38#*
30-39 years	9 (21,0)	3 (17,6)	17 (29,3)	29 (24,6)#
CT/NON-CT	*7/2*	*0/3*	*5/12*	*12/17#*
> 40 years	- (-)	2 (11,8)	3 (5,2)	5 (4,2)#
CT/NON-CT	*-/-*	*1/1*	*0/3*	*1/4#*
TOTAL	43 (100)	17 (100)	58 (100)	118 (100)#
CT/NON-CT	*27/16*	*8/9*	*18/40*	*53/65#*

CT/NON-CT- Cape Town Area/Outside of Cape Town, # - p< 0,01

Table V.2.4 MATERNAL EDUCATIONAL STANDARDS

MATERNAL EDUCATION	BIRTH ORIGIN GREATER CAPE TOWN	OUTSIDE CAPE TOWN	TOTAL
	n (%)	n (%)	n (%)
No schooling	6 (11,3)	24 (36,9)	30 (25,4) #
Primary Schooling only	12 (22,6)	19 (29,2)	31 (26,3) #
Secondary Schooling but no matric	17 (32,1)	12 (18,5)	29 (24,6)#
Matriculated	15 (28,3)	9 (13,9)	24 (20,3)#
Tertiary	3 (5,7)	1 (1,5)	4 (3,4)#
TOTAL	53 (100)	65 (100)	118 (100)

- p < 0,01

The educational profile of the fathers is similar (Table V.2.5).

Table V.2.5 PATERNAL EDUCATIONAL STANDARDS

PATERNAL EDUCATION	BIRTH ORIGIN		TOTAL
	GREATER CAPE TOWN	OUTSIDE CAPE TOWN	
	n (%)	n (%)	n (%)
No schooling	9 (17,0)	24 (36,9)	33 (28,0) #
Primary Schooling only	7 (13,2)	17 (26,2)	24 (20,3) #
Secondary Schooling but no matric	21 (39,6)	13 (20,0)	34 (28,8) #
Matriculated	13 (24,5)	11 (16,9)	24 (20,3) #
Tertiary	3 (5,7)	- (-)	3 (2,5) #
TOTAL	53 (100)	65 (100)	118 (100)

- $p < 0,01$

In two cases there was a family history of a neural tube defect, one being a previously affected pregnancy. An antenatal diagnosis was only made in four pregnancies (3,4%), all by ultrasound examinations. Ninety five mothers (80,5%) had had no antenatal ultrasound examination or any screening for congenital defects during their pregnancies. Nineteen mothers had an antenatal ultrasound examination but no congenital abnormality was reported, eleven were done privately and eight within the Peninsula Maternal and Neonatal Service (Table V.2.6).

Table V.2.6 ANTENATAL DIAGNOSIS

ANTENATAL SCREENING	BIRTH ORIGIN		TOTAL
	GREATER CAPE TOWN	OUTSIDE CAPE TOWN	
	n (%)	n (%)	n (%)
No screening	33 (62,2)	62 (95,4)	95 (80,5)
U/S Private No Diagnosis	9 (17,0)	2 (3,1)	11 (9,3)
U/S PMNS No Diagnosis	8 (15,1)	- (-)	8 (6,8)
U/S PMNS/Private Diagnosis Made	3 (5,7)	1 (1,5)	4 (3,4)
Alpha Fetoprotein	- (-)	- (-)	- (-)
TOTAL	53 (100)	65 (100)	118 (100)

PMNS - Peninsula Maternal and Neonatal Service

The majority of the affected infants were first born, with a progressive decrease in the number of deliveries with increasing birth order, except for a small increase for third born infants (Table V.2.7).

Table V.2.7 BIRTH ORDER

BIRTH ORDER	BIRTH ORIGIN		TOTAL
	GREATER CAPE TOWN	OUTSIDE CAPE TOWN	
	n (%)	n (%)	n (%)
1	24 (45,3)	20 (30,8)	44 (37,3)
2	9 (17,0)	15 (23,0)	24 (20,3)
3	13 (24,5)	13 (20,0)	26 (22,0)
4	5 (9,4)	5 (7,7)	10 (8,5)
5	- (-)	6 (9,2)	6 (5,1)
6	1 (1,9)	4 (6,2)	5 (4,2)
>6	1 (1,9)	2 (3,1)	3 (2,4)
TOTAL	53 (100)	65 (100)	118 (100)

The majority (85,6%) were delivered under medical supervision, either at a peripheral midwifery clinic or at a midwife obstetric unit (within the Peninsula Maternal and Neonatal Service) or in a hospital. There were eleven unattended home deliveries, all outside of Cape Town, and six deliveries en route to medical care, the majority outside of the greater Cape Town area (Table V.2.8).

Table V.2.8 PLACE OF BIRTH

PLACE OF BIRTH	BIRTH ORIGIN		TOTAL
	GREATER CAPE TOWN	OUTSIDE CAPE TOWN	
	n (%)	n (%)	n (%)
Home/BBA	2 (3,8)	15 (23,1)	17 (14,4)
Clinic/MOU	10 (18,9)	5 (7,7)	15 (12,7)
Hospital	41 (77,3)	45 (69,2)	86 (72,9)
TOTAL	53 (100)	65 (100)	118 (100)

BBA - Born Before Arrival; MOU - Midwife Obstetric Unit

Seventy four (62,7%) of the infants were born by normal vertex delivery and a further 17 (14,4%) were, on history, vertex deliveries but were born before arrival either at home or en route to hospital. The majority of these vertex deliveries occurred outside the greater Cape Town area. The two infants in the greater Cape Town area were born before arrival, en route to either hospital or a midwife obstetric unit. The only two breech deliveries as well as the six deliveries requiring forceps assistance all occurred in the greater Cape Town area. Caesarean sections were performed in 19 cases (16,1%), the majority in the greater Cape Town based hospitals (14 vs 5) (Table V.2.9).

Table V.2.9 MODE OF DELIVERY

MODE OF DELIVERY	BIRTH ORIGIN		TOTAL
	GREATER CAPE TOWN	OUTSIDE CAPE TOWN	
	n (%)	n (%)	n (%)
Born Before Arrival/ Home Delivery	2(3,8)	15(23,1)	17(14,4) #
Normal Vertex Delivery	29(54,7)	45(69,2)	74(62,7)
Assisted Delivery	6(11,3)	- (-)	6(5,1)
Breech Delivery	2(3,8)	- (-)	2(1,7)
Caesarean Section	14(26,4)	5(7,7)	19(16,1) #
TOTAL	53(100)	65(100)	118(100)

- $p < 0,01$

In only nine cases was an infant with a large head circumference at birth ($>90^{th}$ centile) delivered by Caesarean section (Table V.2.10).

Sixteen infants (13,6%) were preterm (<37 weeks gestation) at birth, thirteen of whom were more than 36 weeks gestation and the others 35, 32 and 28 weeks gestation respectively. Fifteen (12,7%) of the infants were growth retarded, eight were underweight and seven were small for their gestation (Table V.2.11.).

Table V.2.10 MODE OF DELIVERY vs HEAD CIRCUMFERENCE AT BIRTH

MODE OF DELIVERY	BIRTH HEAD CIRCUMFERENCE		
	NORMAL OR <10TH CENTILE	>90TH CENTILE	TOTAL
	n (%)	n (%)	n (%)
Born Before Arrival	14 (14,6)	3 (13,6)	17 (14,4)
CT/Non-CT	*2/12*	*0/3*	*2/15*
Normal Vertex Delivery	64 (66,7)	10 (45,5)	74 (62,7)
CT/Non-CT	*27/37*	*2/8*	*29/45*
Assisted Delivery	6 (6,3)	- (-)	6 (5,1)
CT/Non-CT	*6/0*	*-/-*	*6/0*
Breech Delivery	2 (2,0)	- (-)	2 (1,7)
CT/Non-CT	*2/0*	*-/-*	*2,0*
Caesarean Section	10 (10,4)	9 (40,9)	19 (16,1)
CT/Non-CT	*6/4*	*8/1*	*14/5*
TOTAL	96 (100)	22 (100)	118 (100)
CT/Non-CT	*43/53*	*10/12*	*53/65*

CT/Non-CT - Cape Town/Non-Cape Town

Table V.2.11 GROWTH PARAMETERS AT BIRTH

GROWTH CLASSIFICATION	BIRTH ORIGIN		
	GREATER CAPE TOWN	OUTSIDE CAPE TOWN	TOTAL
	n (%)	n (%)	n (%)
Gestation <37 Weeks	7 (13,2)	9 (13,8)	16 (13,6)
Appropriate for Gestation	42 (79,2)	48 (73, 8)	90 (76,3)
Large for Gestation	6 (11,3)	7 (10,8)	13 (11,0)
Small for Gestation	5 (9,4)	10 (15,4)	15 (12,7)
COH >90th Centile	10 (18,8)	12 (18,4)	22 (18,7)
2cm or more >90th	5(9,4)	1 (1,5)	6 (5,1)
COH <10th Centile	7 (13,2)	4 (6,2)	11 (9,3)
TOTAL	53 (100)	65 (100)	118 (100)

COH - Head Circumference

Ninety (76,3%) were appropriately grown and 13 (11%) were proportionally large for their gestation. Twenty-two infants had a birth head circumference above the 90th centile (Lubchenco), but in 9 (7,6%) macrocephally was present, six of whom had a head circumference of 2 cms or more above the 90th centile (five from the greater Cape Town area). Eleven infants (9,3%) had a head circumference below the 10th centile at birth.

The anatomical site of the myelomeningocele was lumbar, lumbo-sacral or sacral in 84,8% of the cases (Table V.2.12)

Table V.2.12 ANATOMICAL SITE OF MYELOMENINGOCELE

SITE OF MYELOMENINGOCELE	BIRTH ORIGIN		TOTAL
	GREATER CAPE TOWN	OUTSIDE CAPE TOWN	
	n (%)	n (%)	n (%)
Cervical	- (-)	1 (1,5)	1 (0,8)
Thoracic	1 (1,9)	3 (4,6)	4 (3,4)
Thoraco-Lumbar	6(11,3)	7(10,8)	13(11,0)
Lumbar	14 (26,4)	23 (35,4)	37 (31,4)
Lumbo-Sacral	26 (49,1)	25 (38,5)	51 (43,2)
Sacral	6 (11,3)	6 (9,2)	12 (10,2)
TOTAL	53 (100)	65 (100)	118 (100)

and was similar irrespective of birth origin. The only cervical myelomeningocele was referred from outside of the greater Cape Town area. A sensori-motor level above and including L2 was present in twenty two (18,6%) of the infants, twenty five (21,2%) had mid-lumbar (L3, L4) and L5 sensori-motor levels, respectively, and forty six (39%) had sacral sensori-motor levels (Table V.2.13). Eleven patients had asymmetrical levels differing by no more than one level and were categorised according to the more distal level.

Table V. 2.13 SENSORI-MOTOR LEVEL AT BIRTH

SENSORI-MOTOR LEVEL	BIRTH ORIGIN		TOTAL
	GREATER CAPE TOWN	OUTSIDE CAPE TOWN	
	n (%)	n (%)	n (%)
THORACIC	3 (5,7)	4 (6,2)	7 (5,9)
LUMBAR 1 & 2	6 (11,3)	9 (13,8)	15 (12,7)
LUMBAR 3 & 4	9 (17,0)	16 (24,6)	25 (21,2)
LUMBAR 5	7 (13,2)	18 (27,7)	25 (21,2)
LUMBAR 1 - 5	22 (33,8)	43 (66,2)	65 (55,1)#
SACRAL 1 & 2	13 (24,5)	10 (15,4)	23 (19,5)
SACRAL 3, 4 & 5	15 (28,3)	8 (12,3)	23 (19,5)
ABOVE LUMBAR 5	18 (34,0)	29 (44,6)	47 (39,8) *
LUMBAR 5 & BELOW	35 (66,0)	36 (55,4)	71 (60,2)*
TOTAL	53 (100)	65 (100)	118 (100)

* -Not Significant; # - $p < 0,05$ (for birth origin)

Although lumbar defects were more common amongst infants who were from outside of the greater Cape Town area, the difference for those with a sensori-motor level above L5

was not significant. The correlation coefficient of the sensori-motor levels at birth and the anatomical site of the myelomeningocele was 0,65, with 95% confidence intervals of 0,53 - 0,74. In seventy infants (56%) the myelomeningocele was the only congenital anomaly. In the remainder, other than the myelomeningocele, twenty three infants (18,4%) had congenital talipes equino-varus, 12 (9,6%) had a strabismus, nine (7,2%) had a congenital dislocation of the hip, four (3,2%) had a kyphoscoliosis and seven (5,6%) had other anomalies (Table V.2.14) which ranged from major (a ventricular septal defect requiring surgical correction) to minor anomalies (extra digits) (Table V.2.15). The renal anomalies found were secondary to a neurogenic bladder and are reported with the achievement of continence.

Table V.2.14 CONGENITAL ANOMALIES/DEFORMATIONS

CONGENITAL ANOMALIES	BIRTH ORIGIN		TOTAL
	GREATER CAPE TOWN	OUTSIDE CAPE TOWN	
	n (%)	n (%)	n (%)
M.M.C. ALONE	28 (47,4)	42 (63,6)	70 (56,0)
STRABISMUS	6 (10,2)	6 (9,1)	12 (9,6)
C.D.H.	6 (10,2)	3 (4,6)	9 (7,2)
C.T.E.V.	13 (22,0)	10 (15,1)	23 (18,4)
KYPHO-SCOLIOSIS	3 (5,1)	1 (1,5)	4 (3,2)
OTHER	3 (5,1)	4 (6,1)	7 (5,6)
TOTAL	59(100)*	66 (100)*	125 (100)*

M.M.C. - Myelomeningocele; C.D.H. - Congenital Dislocation of the Hip;
(C.T.E.V) Congenital Talipes Equino-Varus; * - 5 infants had more than one Anomaly

Table V.2.15 OTHER CONGENITAL ANOMALIES

PATIENT	OTHER CONGENITAL ANOMALY	OUTCOME
R.S.	Ventricular Septal Defect	Surgical Correction at 2 years of age
R.T.	Extra Digits bilaterally	Ligated at 2 days of age
M.S.	Cryporchidism	Surgical Correction at 2 years of age
N.H.	Ventricular Septal Defect	Closed spontaneously
F.K.	Hypertrophic Pyloric Stenosis	Pyloromyotomy at 1 month of age
N.B.	Flexion Deformity of Right Knee	Partial resolution with physiotherapy
C.B.	Hypospadias	Surgical Correction at 4 years of age

Other than myelomeningocele, the anomalies appear to be more commonly associated with a mid-lumbar to upper sacral (S1,2) sensori-motor level, with congenital talipes equino-

varus more commonly associated with upper lumbar and thoracic sensori-motor levels, and this difference becomes significant in comparison to the other anomalies mentioned when the sensori-motor levels are grouped into thoracic and upper lumbar (L1,2), mid-lumbar (L3,4), and lower lumbar (L5) and sacral (Table V.2.16).

Table V.2.16 OTHER CONGENITAL ANOMALIES / DEFORMATIONS VS. SENSORI-MOTOR LEVEL

CONGENITAL ANOMALIES	SENSORI-MOTOR LEVEL						TOTAL
	T	L1-2	L3-4	L5	S1-2	S3-5	n (%)
	n	n	n	n	n	n	
STRABISMUS	-	-	2	6	4	-	12 (21,8)
C.D.H.	-	1	5	1	2	-	9 (16,4)
C.T.E.V.	2	5	5	5	4	2	23 (41,8)*
KYPHO-SCOLIOSIS	-	-	2	1	1	-	4 (7,3)
OTHER	-	1	2	1	1	2	7 (12,7)
TOTAL	2	7	16	14	12	4	55 (100)

C.D.H. Congenital Dislocation of the Hip; C.T.E.V. - Congenital Talipes Equino-Varus; * - p < 0,05

Only forty (33,9%) infants were referred to a neurosurgeon and their defect closed within twenty four hours of birth (Table V.2.17).

Table V.2.17 DELAY IN NEUROSURGERY

DELAY IN SURGERY	BIRTH ORIGIN		TOTAL
	GREATER CAPE TOWN (n-53)	OUTSIDE CAPE TOWN (n-65)	(n-118)
	n (%)	n (%)	n (%)
<24HOURS	34(85,0)	6(15,0)	40(33,9)*
>24 HOURS	19(24,4)	59(75,6)	78(66,1)*
<48 HOURS	44 (74,6)	15(25,4)	59(50,0)*
>48 HOURS	9(15,3)	50(84,7)	59(50,0)*
<72 HOURS	44(68,8)	20(31,2)	64(54,2)*
>72 HOURS	9(16,7)	45(83,3)	54(48,8)*
<1 WEEK	47(61,0)	30(38,0)	77 (65,3)*
>1 WEEK	6(14,6)	35(85,4)	41(34,7)*
<2 WEEKS	48(51,6)	45(48,4)	93(78,8)**
> 2 WEEKS	5(20,0)	20(80,0)	25 (21,2)**
<3 WEEKS	49(49,5)	50(50,5)	99(83,9)***
>3 WEEKS	4(21,1)	15(78,9)	19(16,1)***
<4 WEEKS	49(47,6)	52(53,4)	103(87,3)
> 4 WEEKS	4(26,7)	11(73,3)	15(12,7)

* - p <0,001; ** p - <0,01; *** - p< 0,05

Of these the majority (85%) were from the greater Cape Town area. Fifty nine (50%) infants had been referred within 48 hours of birth, forty four (74,6%) from the greater Cape Town area. The number referred progressively increased with time, 103 (87,3%) having been referred within four weeks of birth.

The delays in referral were either as a result of infants being treated conservatively initially, and having survived were then referred, or were transport related in the country cases.

The presence of culture proven infection appears to be related to the delay in treatment. Fifty one infants (86,4%) who were treated within 48 hours of birth were uninfected. The presence of superficial skin infection of the defect itself progressively increased with delay in treatment, reaching a maximum at three weeks of age when twenty one of the ninety nine infants (21,1%) were infected. Culture positive infection of the central nervous system was seen in seventeen infants. Of these five occurred in infants treated within twenty four hours and a further one within forty eight hours of birth, and all these developed subsequent to treatment. With further delays in treatment there is a progressive increase in the number of infants with central nervous system infections, a further nine occurring in infants receiving treatment within four weeks of birth (Table V.2.18).

Ninety one (77,1%) infants had hydrocephalus requiring some form of drainage procedure, one of whom needed an Arnold Chiari decompression procedure. Fifty (42,3%) required only a single ventriculo-peritoneal shunt, twenty nine (24,6%) more than one ventriculo-peritoneal shunt and eleven (9,3%) a ventricular drain and a ventriculo-peritoneal shunt, either before and/or after the drain (Table V.2.19).

Table V.2.18 DELAY IN NEUROSURGERY vs CULTURE PROVEN INFECTION

DELAY IN SURGERY	NO INFECT (n-75)	SUPERFICIAL SKIN INFECT (n-26)	CNS INFECT (n-17)	TOTAL (n-118)
	n (%)	n (%)	n (%)	n (%)
<24 HOURS	34 (85,0)	1 (2,5)*	5 (12,5)	40 (33,9)
>24 HOURS	41 (52,6)	25 (32,0)*	12 (15,4)	78 (66,1)
<48 HOURS	51 (86,4)	2 (3,4)*	6 (10,2)**	59 (50,0)
>48 HOURS	24 (40,7)	24 (40,7)*	11 (18,6)**	59 (50,0)
<72 HOURS	53 (82,8)	5 (7,8)*	6 (9,4)**	64 (54,2)
>72 HOURS	22 (40,7)	21 (38,9)*	11 (20,4)**	54 (45,8)
<1 WEEK	58 (75,3)	10 (13,0)*	9 (11,7)**	77 (65,3)
>1 WEEK	17 (41,5)	16 (39,0)*	8 (19,5)**	41 (34,7)
<2 WEEKS	65 (69,9)	18 (19,4)**	10 (10,7)**	93 (78,8)
> 2 WEEKS	10 (40,0)	8 (32,0)**	7 (28,0)**	25 (21,2)
<3 WEEKS	67 (67,7)	21 (21,2)	11 (11,1)	99 (83,9)
>3 WEEKS	8 (42,1)	5 (26,3)	6 (31,6)	19 (16,1)
<4 WEEKS	67 (65,0)	21 (20,4)	15 (14,6)	103 (67,3)
> 4 WEEKS	8 (53,3)	5 (33,3)	2 (13,3)	15 (12,7)

INFECT. - INFECTION; * - p < 0,001; ** - p< 0,05

Table V.2.19 HYDROCEPHALUS MANAGEMENT

MANAGEMENT	BIRTH ORIGIN		TOTAL
	GREATER CAPE TOWN	OUTSIDE CAPE TOWN	
	n (%)	n (%)	n (%)
NO VP SHUNT	11 (20,8)	16 (24,6)	27 (22,9)
1 VP SHUNT	19 (35,8)	31 (47,7)	50 (42,3)
> 1 VP SHUNT	17 (32,1)	12 (18,5)	29 (24,6)
DRAIN + VP SHUNT	6 (11,3)	5 (7,7)	11 (9,3)
ARNOLD CHIARI DECOMPRESSION	0 (-)	1 (1,5)	1 (0,8)
TOTAL	53 (100)	65 (100)	118 (100)

VP - Ventriculo-Peritoneal

Hydrocephalus requiring surgical treatment was more common amongst infants with a sensori-motor level in the lumbar or thoracic region (Table V.2.20)

Table V.2.20 SENSORI-MOTOR LEVEL AT BIRTH VS HYDROCEPHALUS

SENSORI-MOTOR LEVEL	BIRTH ORIGIN		TOTAL
	NO SHUNT REQUIRED	SHUNT/DRAIN REQUIRED	
	n (%)	n (%)	n (%)
THORACIC	1 (3,7)	6 (6,6)	7 (5,9)*#
LUMBAR 1 & 2	2 (7,4)	13 (14,3)	15 (12,7)*
LUMBAR 3 & 4	3 (11,1)	22 (24,2)	25 (21,2)*
LUMBAR 5	3 (11,1)	22 (24,2)	25 (21,2)*
LUMBAR 1 - 5	8 (29,6)	57 (62,6)	65 (55,1)#
SACRAL 1 & 2	5 (18,5)	18 (19,8)	23 (19,5)*
SACRAL 3,4 & 5	13 (48,1)	10 (11,0)	23 (19,5)*
SACRAL 1- 5	18 (66,7)	28 (30,8)	46 (39 ,0)#
TOTAL	27 (100)	91 (100)	118 (100)

* & # - p < 0,01

as well as a head circumference at birth above the 90th centile on the Lubchenco chart (Table V.2.21).

Table V.2.21 HEAD CIRCUMFERENCE AT BIRTH VC HYDROCEPHALUS

HEAD CIRCUMFERENCE CLASSIFICATION	BIRTH ORIGIN		TOTAL
	NO SHUNT REQUIRED	SHUNT/DRAIN REQUIRED	
	n (%)	n (%)	n (%)
< 10th CENTILE	3(11,1)	8(8,8)	11(9,3)
> 10th < 90th CENTILE	23(85,2)	62(68,1)	85(72,0)
< 90th CENTILE	26(96,3)	70(76,9)	96(81,3)#
> 90th CENTILE	1(3,7)	21(23,1))	22(18,6)#
TOTAL	27(100)	91(100)	118(100)

- p < 0,05

All initial ventriculo-peritoneal shunts were inserted within the first year of life and within eight months of closing the defect. Seventeen (19,5%) of the initial ventriculo-peritoneal shunt placements took place within 7 days and a further twenty (23%) within 14 days of closing the defect closure. Twenty one (24,1%) had over a months delay in shunt placement, the majority being placed within six months, with only two being placed seven months after closure of the defect. (Table V.2.22).

Table V.2.22 DELAY IN INITIAL VP SHUNT PLACEMENT

DELAY IN VP SHUNT	BIRTH ORIGIN		TOTAL
	GREATER CAPE TOWN	OUTSIDE CAPE TOWN	
	n (%)	n (%)	n (%)
< 7 days	5(12,2)	12(26,1)	17(19,5)
> 7 < 14 days	10(24,4)	10(21,7)	20(23,0)
> 14 < 21 days	12(29,3)	9(19,6)	21(24,1)
> 21 < 28 days	5(12,2)	3(6,5)	8(9,2)
> 28 < 56 days	2(4,9)	7(15,2)	9(10,3)
> 56 days	7(17,1)	5(10,9)	12(13,8)
TOTAL	41(100)	46(100)	87(100)

VP - Ventriculo - Peritoneal

All but six of the ventriculo-peritoneal shunt revisions took place in the first year of life. Three of these had occurred before two years of age, two before three years of age and one before five years of age.

Following surgery six died, five within the first week and one forty days post surgery. Three of the deaths were secondary to infection involving the central nervous system. The other three deaths were due to hyaline membrane disease, necrotizing enterocolitis and raised intracranial pressure respectively. Of the three deaths following infection only one was treated within forty eight hours of birth, the other two being treated 8 and 23 days post delivery (Table V.2.23).

Table V.2.23 POST SURGICAL DEATHS

PATIENT	DELAY IN SURGERY	AGE AT DEATH	CAUSE OF DEATH
R.T.	< 24 hr	2 days	Hyaline Membrane Disease/Pulmonary Haemorrhage
T.C.	< 24 hr	3 days	Recurrent Apnoea/Raised Intracranial Pressure
N.N.	< 24 hr	3 days	Necrotizing Enterocolitis
M.V.	< 48 hr	40 days	Ventriculitis
B.M.	8 days	9 days	Ventriculitis
B.S.	23 days	28 days	Septicaemia/Ventriculitis

Six infants died subsequently, three within the first year and three within the second year of life. All those who died in the first year of life died of infection, one involving the

central nervous system and the other two both had a septicaemia one following gastro-enteritis and the other a bronchopneumonia. Two of the infants who died in their second year of life, died of a septicaemia following a pneumonic illness, one complicating chronic renal failure. The other infant died of raised intracranial pressure following blockage of his ventriculo-peritoneal shunt (Table V.2.24).

Table V.2.24 SUBSEQUENT DEATHS

PATIENT	DELAY IN SURGERY	AGE AT DEATH	CAUSE OF DEATH
M.H.	< 24 hr	6 mths	Septicaemia/Gastro-enteritis
P.D.	< 24 hr	4 mths	Septicemia/Gastro-enteritis
P.v.N.	< 24 hr	3 mths	Septicaemia/Pneumonia
R.v.R.	< 24 hr	2 years	Pneumonia/Chronic Renal Failure
Z.K.	< 24 hr	1 y 4 m	Septicaemia/Pneumonia
J.P.	5 days	1 y 1 m	Raised Intracranial/Pressure Blocked VPS

VPS - Ventriculo-Peritoneal Shunt

Fourteen infants were lost to follow-up, 8 in the first year (4B, 4MA; 1 from Cape Town, and 7 from > 100 kms from Cape Town), 3 in the second year (1 B, 2 MA; all >100 kms from Cape Town) and 3 in the 4th year of life (3B; all > 500 kms from Cape Town). Twelve infants are being followed-up elsewhere, 7 in Kimberley and 5 in East London. Eighty nine infants were seen at one year, eighty four infants at two and three years, and eighty infants at four and five years of age, respectively.

On testing with the rapid developmental assessment used at the spinal defects clinic, the mean developmental quotient(DQ) was 89,31 with a standard deviation of 15,83 at one, 86,08 with a standard deviation of 18,24 at two, 84,88 with a standard deviation of 17,62 at three, and 85,09 with a standard deviation of 18,53 at four years of age. At five years of age, testing with the Griffiths Scales of Mental Development the mean General Quotient (GQ) was 76 with a standard deviation of 16,91. The mean modified GQ (excluding the locomotor scale) was 80,44 with a standard deviation of 17,56 (Table V.2.25).

Table V.2.25 DEVELOPMENTAL/GENERAL QUOTIENTS VS AGE

	INFANT AGE IN YEARS (YRS)				
	1 Yr	2 Yr	3 Yr	4 Yr	5 Yr
No.	89 (100)	84 (100)	84 (100)	80 (100)	80 (100)
Mean DQ/GQ	89,31	86,06	84,88	85,09	76,0 80,44 #
SD	15,83	18,24	17,62	18,53	16,91 17,56 #

- Modified GQ; GQ - General Quotient; DQ - Developmental Quotient; SD Standard Deviation

Sixty nine infants (77,5%) had a DQ above eighty, and only two (2,2%) had a DQ less than fifty at one year of age. Sixty one (72,6%) had a DQ above eighty, and only five (6%) had a DQ less than 50 at two years of age. Fifty eight (69%) had a DQ above eighty, and only four (4,8%) had a DQ less than 50 at three years of age. Fifty seven (71,3%) had a DQ above eighty, and only four (4,8%) had a DQ less than 50. Thirty nine (48,8%) had a GQ and forty nine (61,3%) had a modified GQ above eighty, and only six (7,5%) had a GQ and five (6,3%) had a modified GQ less than 50 at five years of age (Table V.2.26).

Table V.2.26 DEVELOPMENTAL/GENERAL QUOTIENTS VS AGE

	INFANT AGE IN YEARS (YRS)				
DQ/GQ CATEGORY	1 Yr	2 Yr	3 Yr	4 Yr	5 Yr
	n (%)	n (%)	n (%)	n (%)	n (%)
NORMAL (> 80)	69 (77,5)	61 (72,6)	58 (69,0)	57 (71,3)	39 (48,8) 49 (61,3) #
BORDERLINE (70 - 79)	11 (12,4)	8 (9,5)	11 (13,1)	5 (6,3)	15 (18,8) 11 (13,8) #
MILD (50 - 69)*	7 (7,9)	10 (11,9)	11 (13,1)	14 (17,5)	20 (25,0) 15 (18,8) #
MODERATE (30 - 49)*	2 (2,2)	5 (6,0)	4 (4,8)	3 (3,8)	6 (7,5) 5 (6,3) #
SEVERE (< 30)*	- (-)	- (-)	- (-)	1 (1,3)	- (-) (-) #
TOTAL	89 (100)	84 (100)	84 (100)	80 (100)	80 (100)

- Modified GQ (MGQ) * - Retardation

Of the variables compared to the DQ (Tables V.2.27, V.2.28,V.2.V9,V.2.30)

Table V.2.27 DEVELOPMENTAL QUOTIENTS AT 1 YEAR OF AGE

	DEVELOPMENTAL QUOTIENTS					
	n	Mean	S.D.	n	Mean	S.D
BIRTH ORIGIN						
CT/Outside CT	45	88	15	44	90	17
DELAY IN NEUROSURGERY						
< 24 Hr/ > 24 Hr	31	93	14	58	88	17
< 48 Hr/ > 48 Hr	48	91	15	41	87	17
<72 Hr/ >72 Hr	52	90	16	28	88	16
< 1 WEEK/ > 1 WEEK	61	90	16	28	88	16
INFECT./NO INFECT.	12	81	16	77	91	16 #
SHUNT STATUS						
No Shunt /1 Shunt	20	93	9	37	92	15
No Shunt/ >1 Shunt	20	93	9	32	84	18 #
1 Shunt/ > 1 Shunt	37	92	15	32	84	18 #

- $p < 0,05$

Table V.2.28 DEVELOPMENTAL QUOTIENTS AT 2 YEARS OF AGE

	DEVELOPMENTAL QUOTIENTS					
	n	Mean	S.D.	n	Mean	S.D
BIRTH ORIGIN						
CT/Outside CT	40	85	18	44	87	19
DELAY IN NEUROSURGERY						
< 24 Hr/ > 24 Hr	31	89	15	53	84	20
< 48 Hr/ > 48 Hr	47	87	18	37	85	19
<72 Hr/ >72 Hr	50	86	19	34	86	18
< 1 WEEK/ > 1 WEEK	58	86	18	26	86	19
INFECT./NO INFECT.	11	81	22	73	87	18
SHUNT STATUS						
No Shunt /1 Shunt	19	90	13	35	87	17
No Shunt/ >1 Shunt	19	90	13	30	82	22
1 Shunt/ > 1 Shunt	35	87	17	30	82	22

Table V.2.29 DEVELOPMENTAL QUOTIENTS AT 3 YEARS OF AGE

	DEVELOPMENTAL QUOTIENTS					
	n	Mean	S.D.	n	Mean	S.D
BIRTH ORIGIN						
CT/Outside CT	40	85	17	44	85	18
DELAY IN NEUROSURGERY						
< 24 Hr/ > 24 Hr	31	88	15	53	83	19
< 48 Hr/ > 48 Hr	47	87	18	37	83	17
<72 Hr/ >72 Hr	50	86	18	34	83	17
< 1 WEEK/ > 1 WEEK	58	85	18	26	84	16
INFECT./NO INFECT.	11	77	20	73	86	17
SHUNT STATUS						
No Shunt /1 Shunt	19	87	14	35	88	17
No Shunt/ >1 Shunt	19	87	14	30	80	20
1 Shunt/ > 1 Shunt	35	88	17	30	80	20

Table V.2.30 DEVELOPMENTAL QUOTIENTS AT 4 YEARS OF AGE

	DEVELOPMENTAL QUOTIENTS					
	n	Mean	S.D.	n	Mean	S.D
BIRTH ORIGIN						
CT/Outside CT	36	84	19	44	87	19
DELAY IN NEUROSURGERY						
< 24 Hr/ > 24 Hr	31	88	16	49	83	20
< 48 Hr/ > 48 Hr	47	87	18	33	83	19
<72 Hr/ >72 Hr	50	86	18	30	83	19
< 1 WEEK/ > 1 WEEK	58	85	19	22	85	19
INFECT./NO INFECT.	11	73	26	69	87	17 #
SHUNT STATUS						
No Shunt /1 Shunt	19	90	13	31	90	17
No Shunt/ >1 Shunt	19	90	13	30	77	20 #
1 Shunt/ > 1 Shunt	31	90	17	30	77	20 #

- $p < 0,05$

and GQ and modified GQ (Tables V.2.31 and V.2.32)

Table V.2.31 GENERAL QUOTIENTS AT 5 YEARS OF AGE

	GENERAL QUOTIENTS					
	n	Mean	S.D.	n	Mean	S.D
BIRTH ORIGIN						
CT/Outside CT	36	74	15	44	78	18
DELAY IN NEUROSURGERY						
< 24 Hr/ > 24 Hr	31	80	15	49	73	18
< 48 Hr/ > 48 Hr	47	79	17	33	72	17
<72 Hr/ >72 Hr	50	78	17	30	73	16
<1 WEEK/ > 1 WEEK	58	77	17	22	73	16
INFECT./NO INFECT.	11	67	17	69	77	17#
SHUNT STATUS						
No Shunt /1 Shunt	19	80	12	31	80	17
No Shunt/ >1 Shunt	19	80	12	30	69	18#
1 Shunt/ > 1 Shunt	31	80	17	30	69	18#

- $p < 0,05$

Table V.2.32 MODIFIED GENERAL QUOTIENTS AT 5 YEARS OF AGE

	GENERAL QUOTIENTS					
	n	Mean	S.D.	n	Mean	S.D
BIRTH ORIGIN						
CT/Outside CT	36	79	15	44	81	20
DELAY IN NEUROSURGERY						
< 24 Hr/ > 24 Hr	31	84	16	49	78	18
< 48 Hr/ > 48 Hr	47	83	18	33	77	17
<72 Hr/ >72 Hr	50	82	18	30	78	16
<1 WEEK/ > 1 WEEK	58	81	18	22	79	16
INFECT./NO INFECT.	11	71	18	69	82	17#
SHUNT STATUS						
No Shunt /1 Shunt	19	83	13	31	85	17
No Shunt/ >1 Shunt	19	83	13	30	74	19#
1 Shunt/ > 1 Shunt	31	85	17	30	74	19#

- $p < 0,05$

significant differences were evident for the presence of culture positive infection, and the placement of more than one ventriculoperitoneal shunt at one, four and five years of age. A difference in GQ and each of the subtests was found between those who required repeated shunting (two or more shunts) and those shunted once (Table V.2.33).

Table V.2.33 EFFECTS OF SHUNTING ON SUB-SCALE QUOTIENTS AT 5 YEARS

GRIFFITHS SUB -SCALES	GENERAL QUOTIENTS					
	Not Shunted (n-19)		One Shunt (n-31)		More than One Shunt (n-30)	
	Mean	S.D.	Mean	S.D.	Mean	S.D.
Locomotion	67	26	62	27	43	29 #
Personal/Social	87	13	86	17	76	16 #
Hearing/Speech	86	15	88	17	79	23 #
Eye/Hand Co-ordination	85	14	83	15	74	21 #
Performance	81	15	83	16	72	20 #
Practical Reasoning	79	15	81	30	70	22 #

- $p < 0,05$ for Not Shunted vs More than One Shunt and for One Shunt vs More than One Shunt

Similarly, differences were demonstrated between the group that escaped shunting and those infants shunted more than once. The infants who sustained central nervous system infection performed less well than those uninfected on each of the Griffiths subtests (Table V.2.34)

Table V.2.34 EFFECTS OF INFECTION ON SUB-SCALE QUOTIENTS AT 5 Yr

GRIFFITHS SUB -SCALES	GENERAL QUOTIENTS			
	NOT INFECTED (n-69)		INFECTED (n-11)	
	Mean	S.D.	Mean	S.D.
Locomotion	56	29	46	28 #
Personal/Social	83	16	74	20 #
Hearing/Speech	87	19	72	22 #
Eye/Hand Co-ordination	82	18	68	18 #
Performance	80	18	68	21 #
Practical Reasoning	79	21	66	22 #

- $p < 0,05$

Of the sixty one infants who received shunts, eleven developed intracranial infections. These children scored lower on the modified GQ (81,46 vs 70,10; t=1,787; p 0,05) with significant differences on the Hearing/Speech and Eye/Hand subtests. When the children with shunts and no infection were compared to those not shunted without infection, no significant differences were found. Taking the cohort as a whole, the lowest subtest quotient was locomotion. The remaining subtests were similar, although slightly higher

quotients were obtained in personal/social and hearing/speech compared to the performance and practical reasoning domains.

The DQ obtained from summing the subtests of the rapid screening developmental assessment (DEI) used at the Spinal Defects Clinic, correlated at one, two, three and four years. The correlation's were found to improve over the four years (Table V.2.35).

Table V.2.35 CORRELATION OF DEI AND GRIFFITHS SCALES

	CORRELATION COEFFICIENT	95% CONFIDENCE INTERVAL
DEI at 1 YEAR	0,77	0,66 - 0,85
DEI at 2 YEARS	0,83	0,75 - 0,89
DEI at 3 YEARS	0,84	0,77 - 0,90
DEI at 4 YEARS	0,89	0,84 - 0,93

Lumbosacral sensori-motor levels were present in eighty five (95,5%) of the infants at one year, eighty (95,2%) at two and three years, and seventy six (95%) had a lumbosacral sensori-motor level (Table V.2.36).

Nine infants (10,1%) were ambulant at one, twenty eight infants (33,3%) at two, forty three infants (51,2%) at three, fifty five infants (68,8%) at four, and fifty seven infants (71,3%) at five years of age. Of the ambulant infants all had a sacral sensori-motor level at one year of age. At two, three, four and five years of age, all a sensori-motor level below L3. Of the ambulant infants with a lumbar sensori-motor level, two were mid-lumbar (L3 or L4) and one lower lumbar (L5) at two, four were midlumbar and five lower lumbar at three, twelve midlumbar and seven lower lumbar at four, and fourteen midlumbar and seven lower lumbar at five years of age, respectively (Table V.2.36). Compared to the anatomical site of the lesion at birth, the ambulant infants all had a lumbosacral or sacral lesion at one year of age, a lumbar, lumbosacral or sacral lesion at two or three years of age, and a thoracolumbar, lumbar, lumbosacral or sacral at four or

five years, respectively (Table V.2.36.1). In comparison to ambulation and sensori-motor level, significance for ambulation and anatomical site of the lesion at birth was only seen

Table V.2.36 AMBULATION VS SENSORI-MOTOR LEVEL

	AMBULATION PER YEAR (Yr) OF AGE					
SENSORI-MOTOR LEVEL	1 Yr n (%)	2 Yr n (%).	3 Yr n (%)	4 Yr n(%)	5 Yr n(%)	
THORACIC	4 (4,5)	4 (4,8)	4 (4,8)	4 (5,0)	4 (5,0)	#
A/NA	*0/4*	*0/4*	*0/4*	*0/4*	*0/4*	
LUMBAR 1-2	9 (10,1)	7 (8,3)	7 (8,3)	6 (7,5)	6 (7,5)	
A/NA	*0/9*	*0/7*	*0/7*	*0/6*	*0/6*	
LUMBAR 3-4	31 (34,8)	30 (35,8)	30 (35,8)	27 (33,8)	27 (33,8)	#
A/NA	*0/31*	*2/28*	*4/26*	*12/15*	*14/13*	
LUMBAR 5	7 (7,9)	7 (8,3)	7 (8,3)	7 (8,6)	7(8,6)	
A/NA	*0/7*	*1/6*	*5/2*	*7/0*	*7/0*	
SACRAL	38 (42,7)	36(42,8)	36 (42,8)	36 (45,0)	36 (45,0)	#
A/NA	*9/29*	*25/11*	*34/2*	*36/0*	*36/0*	
TOTAL	89 (100)	84 (100)	84 (100)	80 (100)	80 (100)	
A/NA	*9/80*	*28/56*	*43/41*	*55/25*	*57/23*	

# p < 0,001 for 1 Year, p < 0,0001 for 2-5 years *A/NA* - Ambulant/ Not Ambulant

for lumbar, lumbosacral and sacral lesions.

Table V.2.36.1 AMBULATION VS ANATOMICAL SITE OF LESION

	AMBULATION PER YEAR (Yr) OF AGE					
MYELOMENING. SITE	1 Yr n (%)	2 Yr n (%).	3 Yr n (%)	4 Yr n(%)	5 Yr n(%)	
CERVICAL	1 (1,1)	1 (1,2)	1 (1,2)	1 (1,2)	1 (1,2)	
A/NA	*0/1*	*0/1*	*0/1*	*0/1*	*0/1*	
THORACIC	2 (2,3)	2 (2,4)	2 (2,4)	2 (2,5)	2 (2,5)	
A/NA	*0/2*	*0/2*	*0/2*	*0/2*	*0/2*	
THORACOLUMBAR	9 (10,1)	9 (10,7)	9 (10,7)	8 (10,0)	8 (10,0)	
A/NA	*0/9*	*0/9*	*0/9*	*1/7*	*2/6*	
LUMBAR	27 (30,3)	25(29,8)	25(29,8)	22(27,5)	22(27,5)	!
A/NA	*0/27*	*6/20*	*9/16*	*15/7*	*16/6*	
LUMBOSACRAL	40 (44,9)	37 (44,0)	37 (44,0)	37 (46,3)	37 (46,3)	*
A/NA	*4/36*	*14/23*	*25/12*	*30/7*	*30/7*	
SACRAL	10 (11,3)	10(11,9)	10 (11,9)	10 (12,5)	10 (12,5)	#
A/NA	*5/5*	*8/2*	*9/1*	*3/1*	*9/1*	
TOTAL	89 (100)	84 (100)	84 (100)	80 (100)	80 (100)	
A/NA	*9/80*	*28/56*	*43/41*	*55/25*	*57/23*	

- p < 0,001 for 1 Yr., p < 0,05 for 2&3 Yrs.;

* - p<0,001 for 3 Yrs., p<0,01 for 1,2,4&5 Yrs.;

!p<0,001 for 3-5 Yrs., p<0,01 for 2 Yrs; *A/NA* - Ambulant/ Not Ambulant

Of the ambulant infants five (55,5%) at one, nineteen (67,8%) at two, twenty seven (62,8%) at three, thirty four (61,8%) at four, and thirty five (61,45%) at five years of age, were from the greater Cape Town area (Table V.2.37).

Table V.2.37 AMBULATION VS BIRTH ORIGIN

	AMBULATION PER YEAR (Yr.) OF AGE				
BIRTH ORIGIN	1 Yr	2 Yr	3 Yr	4 Yr	5 Yr
	n (%)	n (%)	n (%).	n (%)	n (%)
GREATER CAPE TOWN	44 (49,4)	44 (52,4)	44 (52,4)	44 (55,0)	44 (55,0)
A/NA	*5/39*	*19/25#*	*27/17*	*34/10*	*35/9*
OUTSIDE CAPE TOWN	45 (50,6)	40 (47,6)	40 (47,6)	36 (45,0)	36 (45,0)
A/NA	*4/41*	*9/31#*	*16/24*	*21/15*	*22/14*
TOTAL	89 (100)	84 (100)	84 (100)	80 (100)	80 (100)
A/NA	*9/80*	*28/56#*	*43/41*	*55/25*	*57/23*

- p < 0,05 *A/NA* - Ambulant/Not Ambulant

The number of infants requiring orthopaedic surgery was far higher in those with lumbar sensori-motor levels than thoracic or sacral. However, the highest number requiring orthopaedic surgery occurred in those with mid-lumbar (L3 and L4) and lower lumbar (L5) sensori-motor levels, and for the midlumbar group this reached statistical significance (Table V.2.38).

Table V.2.38 ORTHOPAEDIC SURGERY VS SENSORI-MOTOR LEVEL

	ORTHOPAEDIC SURGERY PER YEAR (Yr.) OF AGE					
SENSORI-MOTOR LEVEL	1 Yr n(%)	2 Yr n(%)	3 Yr n(%)	4 Yr n(%)	5 Yr n(%).	
THORACIC	4 (4,5)	4 (4,8)	4 (4,8)	4 (5,0)	4 (5,0)	#
R/NR	*1/3*	*1/3*	*1/3*	*1/3*	*1/3*	
LUMBAR 1-2	9 (10,1)	7 (8,3)	7 (8,3)	6 (7,5)	6 (7,5)	
R/NR	*4/5*	*4/3*	*4/3*	*3/3*	*3/3*	
LUMBAR 3-4	31 (34,8)	30 (35,8)	30 (35,8)	27 (33,8)	27 (33,8)	#
R/NR	*26/5*	*26/4*	*26/4*	*23/4*	*23/4*	
LUMBAR 5	7 (7,9)	7 (8,3)	7 (8,3)	7 (8,6)	7 (8,6)	
R/NR	*5/2*	*5/2*	*5/2*	*5/2*	*5/2*	
SACRAL	38 (42,7)	36 (42,8)	36 (42,8)	36 (45,0)	36 (45,0)	#
R/NR	*10/28*	*10/26*	*10/26*	*10/26*	*10/26*	
TOTAL	89 (100)	84 (100)	84 (100)	80 (100)	80 (100)	
R/NR	*46/43*	*46/38*	*46/38*	*42/38*	*42/38*	

-p < 0,001 for each year *R/NR*- Required/Not Required

Maternal education did not appear to play a major role in the ambulation or the age at which ambulation occurs (Table V.2.39).

Table V.2.39 AMBULATION VS MATERNAL EDUCATION

MATERNAL EDUCATIONAL STANDARD	INFANT AGE				
	1 Yr	2 Yr.	3 Yr	4 Yr	5 Yr
	n (%)	n (%)	n (%)	n (%)	n (%)
NO SCHOOLING	17 (19,1)	17 (20,2)	17 (17,4)	15 (18,8)	15 (18,8)
A/NA	*1/16*	*4/13*	*7/10*	*8/7*	*9/6*
PRIMARY SCHOOL ONLY	21 (23,8)	19 (22,6)	19 (24,0)	18 (24,5)	18 (22,5)
A/NA	*3/18*	*7/12*	*8/11*	*11/7*	*12/6*
SECONDARY SCHOOL NO MATRIC	25 (28,1)	23 (27,4)	23 (24,0)	22 (27,5)	22 (27,5)
A/NA	*2/23*	*8/15*	*13/10*	*17/5*	*17/5*
MATRICULATED	23 (25,8)	22 (26,2)	22 (29,3)	22 (27,5)	22 (27,5)
A/NA	*3/20*	*8/14*	*14/8*	*17/5*	*17/5*
TERTIARY	3 (3,4)	3 (3,6)	3 (5,3)	3 (3,7)	3 (3,7)
A/NA	*0/3*	*1/2*	*1/2*	*2/1*	*2/1*
TOTAL	89 (100)	84 (100)	84 (100)	80 (100)	80 (100)
A/NA	*9/80*	*28/56*	*43/41*	*55/25*	*57/23*

A/NA - Ambulant/Not Ambulant

Initially social class did not appear to play a role in ambulation (<4 years of age), but far more infants remained ambulant in Social Class I than in Social Class II and III at four and five years of age. This may be a reflection of the fact that more Social Class III families came from outside of Cape Town and required transport to attend clinics and ongoing maintenance of appliances is neglected (Table V.2.40).

Table V.2.40 AMBULATION VS SOCIAL CLASS

INFANT AGE	SOCIAL CLASS			
	I	II	III	TOTAL.
	n (%)	n (%)	n (%)	n (%)
1 YEAR	39 (43,8)	13 (14,6)	37 (41,6)	89 (100,0)
A/NA	*4/35*	*2/11*	*3/34*	*9/80*
2 YEARS	37 (44,0)	12 (14,3)	35 (41.7)	84 (100,0)
A/NA	*14/23*	*4/8*	*10/25*	*28/56*
3 YEARS	37 (44,0)	12 (14,3)	35 (41,7)	84 (100,0)
A/NA	*22/15*	*7/5*	*14/21*	*43/41*
4 YEARS	37 (46,2)	11 (13,8)	32 (40,0)	80 (100,0)
ANA	*30/7*	*8/3*	*17/15*	*55/25**
5 YEARS	37 (46,2)	11 (13,8)	32 (40,0)	80 (100)
A/NA	*19/13*	*0/9*	*6/23*	*25/45**

* $p < 0,05$; *A/NA* - Ambulant/Not Ambulant

The ambulatory status of these infants did appear to be related to their intelligence. Those that were ambulant had a higher developmental or general quotient than those not ambulant and this difference became significant over the age of 2 years and became more so over the age of 4 years. This difference remains significant even when comparing the modified general quotients (excluding the ambulatory subscale) between the two groups (Table V.2.41).

Table V.2.41 AMBULATORY STATUS VS DEVELOPMENTAL/GENERAL QUOTIENTS

ASSESSMENT AGE	AMBULATORY STATUS					
	AMBULANT			NOT AMBULANT		
	n	Mean	S.D.	n	Mean.	S.D.
AT 1 YEAR (DQ)	9	94	9	80	89	16
AT 2 YEARS (DQ)	28	95	12	56	82	19#
AT 3 YEARS (DQ)	43	91	13	41	79	20#
AT 4 YEARS (DQ)	55	91	15	25	72	20*
AT 5 YEARS (DQ)	57	82	14	23	62	16*
AT 5 YEARS (MGQ)	57	85	15	23	69	20*

* - p < 0,001; DQ - Developmental Quotient (DEI);
GQ-General Quotients (Griffiths); MGQ-Modified GQ

Race and sex did not appear to play a major role in ambulation of these infants, except that in the black group infants become ambulant later (Tables V.2.42 and V.2.43)

Table V.2.42 AMBULATORY STATUS VS RACE

RACE	INFANT AGE				
	1 Yr	2 Yr	3 Yr	4 Yr	5 Yr.
	n (%)	n (%)	n (%)	n (%)	n (%)
WHITE	24 (27,0)	22 (26,2)	22(26,2)	22 (27,5)	22 (27,5)
A/NA	*3/21*	*7/15*	*13/9 #*	*16/6*	*16/6*
MIXED ANCESTRY	51(57,3)	49 (58,3)	49 (58,3)	49 (61,3)	49 (61,3)
A/NA	*6/45*	*20/29*	*28/21 #*	*35/14*	*36/13*
BLACK	14 (15,7)	13 (15,5)	13 (15,5)	9 (11,2)	9 (11,2)
A/NA	*0/14*	*1/12*	*2/11 #*	*4/5*	*5/4*
TOTAL	89 (100)	84 (100)	84 (100)	80 (100)	80 (100)
A/NA	*9/80*	*28/56*	*43/41*	55/25	*57/23*

- p < 0,05 *A/NA* - Ambulant/Not Ambulant

Table V.2.43 AMBULATORY STATUS VS SEX

	INFANT AGE				
SEX	1 Yr	2 Yr	3 Yr	4 Yr	5 Yr
	n (%)	n (%)	n (%)	n (%)	n (%)
MALE	48 (53,9)	44 (52,4)	44 (52,4)	44 (55,0)	44 (55,0)
A/NA	*3/45*	*16/28*	*32/12*	*32/12*	*33/11*
FEMALE	41 (46,1)	40 (47,6)	40 (47,6)	36 (45,0)	36 (45,0)
A/NA	*6/35*	*12/28*	*17/23*	*23/13*	*24/12*
TOTAL	89 (100)	84 (100)	84 (100)	80 (100)	80 (100)
A/NA	*9/80*	*28/56*	*43/41*	*55/25*	*57/23*

A/NA - Ambulant/Not Ambulant

Normally urinary continence is only achieved between 2 and 3 years of age. At two years of age one infant with a sensori-motor level of S5 was dry during the day without any assistance. Fifteen (18%) infants at three years of age were dry during the day, nine without assistance and six on a three hourly intermittent transurethral catheterisation programme (Table V.2.44).

Table V.2.44 CONTINENCE AT 3 YEARS VS SENSORI-MOTOR LEVEL

	URINARY CONTINENCE			
SENSORI-MOTOR LEVEL	CONTINENT NO ASSIST.	CONTINENT ON I.C.	NOT CONTINENT	TOTAL
	n (%)	n (%)	n (%)	n (%)
THORACIC	-(-)	1 (16,7)	3 (4,4)	4 (4,8)*
LUMBAR 1-2	- (-)	- (-)	7 (10,1)	7 (8,3)
LUMBAR 3-4	-(-)	2 (33,3)	28 (40,7)	30 (35,8)
LUMBAR 5	-(-)	- (-)	7(10,1)	7 (8,3)
LUMBAR 1-5	-(-)	2 (33,3)	42 (60,9)	44 (52,4)*
SACRAL	9 (100,0)	3 (50,00	24 (34,7)	36 (42,8)*
TOTAL	9 (100,0)	6 (100,0)	69 (100,00	84 (100,0)

- $p < 0,01$; I.C. - Intermittent Catheterisation

Thirty two infants at four and thirty five infants at five years of age were dry during the day, ten without assistance, twenty two at four and twenty five at five years of age were successfully managed on the three hourly intermittent transurethral catheterisation programme. All infants who were continent without assistance had sacral sensori-motor levels, and if those successfully managed on intermittent catheterisation are included, more infants with a sacral sensori-motor level are continent (Table V.2.45 and V.2.46).

Table V.2.45 CONTINENCE AT 4 YEARS VS SENSORI-MOTOR LEVEL

SENSORI-MOTOR LEVEL	URINARY CONTINENCE			
	CONTINENT NO ASSIST.	CONTINENT ON I.C.	NOT CONTINENT	TOTAL
	n (%)	n (%)	n (%)	n (%)
THORACIC	- (-)	3 (13,6)	1 (2,1)	4 (5,0)*
LUMBAR 1-2	- (-)	- (-)	6 (12,5)	6 (7,5)
LUMBAR 3-4	-(-)	7 (31,8)	20 (41,7)	27 (33,8)
LUMBAR 5	- (-)	3 (13,6)	4 (8,3)	7 (8,7)
LUMBAR 1-5	-(-)	10 (45,5)	30 (62,5)	40 (50,0)*
SACRAL	10 (100,0)	9 (40,9)	17 (35,4)	36 (45,0)*
TOTAL	10 (100, 0)	22 (100,0)	48 (100,0)	80 (100,0)

- p < 0,05; I.C. - Intermittent Catheterisation

Table V.2.46 CONTINENCE AT 5 YEARS VS SENSORI-MOTOR LEVEL

SENSORI-MOTOR LEVEL	URINARY CONTINENCE			
	CONTINENT NO ASSIST.	CONTINENT ON I.C.	NOT CONTINENT	TOTAL
	n (%)	n (%)	n (%)	n (%)
THORACIC	- (-)	3 (12,0)	1 (2,2)	4 (5,0)*
LUMBAR 1-2	- (-)	1 (4,0)	5 (11,1)	6 (7,5)
LUMBAR 3-4	- (-)	7 (28,0)	20 (44,4)	27 (33,8)
LUMBAR 5	- (-)	4 (16,0)	3 (6,7)	7 (8,7)
LUMBAR 1-5	- (-)	12 (34,3)	28 (62,2)	40 (50,0)*
SACRAL	10 (100,0)	10 (40,0)	16 (35,6)	36 (45,0)*
TOTAL	10 (100,0)	25 (100,0)	45 (100,0)	80 (100,0)

- p < 0,05; I.C. - Intermittent Catheterisation

To achieve continence using the intermittent catheterisation programme requires ongoing and sometimes frequent medical and nursing support, especially at the commencement of the programme. More infants from the greater Cape Town area were continent than those from outside of Cape Town, and this difference reached significance at 5 years of age (Table V.2.47).

Table V.2.47 CONTINENCE ON I.C. PROGRAMME VS BIRTH ORIGIN

CONTINENCE STATUS	BIRTH ORIGIN		TOTAL
	GREATER CAPE TOWN n (%)	OUTSIDE CAPE TOWN n (%)	n (%)
AT 3 YEARS	37 (49,3)	38 (49,7)	75 (100,0)
CONT/NOT CONT.	*5/32*	*1/37*	*6/69*
AT 4 YEARS	36 (51,4)	34 (48,6)	70 (100,0)
CONT/NOT CONT.	*15/21*	*7/27*	*22/48*
AT 5 YEARS	36 (51,4)	34 (48,6)	70 (100,0)
CONT/NOT CONT.	*17/19*	*8/26*	*25/45**

* - p < 0,05; I.C. - Intermittent Catheterisation; *CONT.* - Continent.

Comparing the developmental and general quotients between continent and incontinent infants, there tends to be a higher quotient in the infants who were continent and this achieves significance at 5 years of age (Table V 2.48).

Table V.2.48 CONTINENCE ON INTERMITTENT CATHETERISATION PROGRAMME VS DEVELOPMENTAL/GENERAL QUOTIENTS

ASSESSMENT AGE	URINARY CONTINENCE					
	CONTINENT ON I.C.			NOT CONTINENT		
	n	Mean	S.D.	n	Mean	S.D.
AT 3 YEARS (DQ)	6	86	18	69	84	18
AT 4 YEARS (DQ)	22	89	17	48	81	19
AT 5 YEARS (GQ)	25	79	15	45	71	17 *
AT 5 YEARS (MGQ)	25	84	16	45	75	17 *

- $p < 0,05$; I.C. - Intermittent Catheterisation DQ - Developmental Quotient (DEI); GQ-General Quotients (Griffiths); MGQ-Modified GQ

Urinary incontinence increased with decreasing social class. More children from social class II and III were incontinent in comparison to social class I, reaching significance at 4 and 5 years of age (Table V.2.49).

Table V.2.49 CONTINENCE ON I.C. PROGRAMME VS SOCIAL CLASS

INFANT AGE	SOCIAL CLASS			
	I	II.	III	TOTAL
	n (%)	n (%)	n (%)	n (%)
3 YEARS	32 (42,7)	10 ((13,3)	33 (44,0)	75 (100,0)
CONT/NOT CONT	*5/27*	*0/10*	*1/32*	*6/69*
4 YEARS	32 (45,7)	9 (12,9)	29 (41,4)	70 (100,0)
CONT/NOT CONT	*18/14*	*0/9*	*4/25*	*22/4**
5 YEARS	32 (45,7)	9 (12,9)	29 (41,4)	70 (100,0)
CONT/NOT CONT	*19/13*	*0/9*	*6/23*	*25/45**

* - $p < 0,001$; I.C. Intermittent Catheterisation; *CONT.* - Continent

As the intermittent catheterisation programme demands full involvement of the mother, it is of note that more infants of mothers who had matriculated were continent at 4 and 5 years of age (Table V.2.50).

Table V.2.50 CONTINENCE ON I.C. PROGRAMME VS MATERNAL EDUCATION

	INFANT AGE		
MATERNAL EDUCATIONAL STANDARD	**3 YEARS**	**4 YEARS.**	**5 YEARS**
	n (%)	**n (%)**	**n (%)**
NO SCHOOLING	13 (17,4)	13 (18,5)	13 (18,5)
Ct/NCt	*0/13*	*1/12**	*1/12**
PRIMARY SCHOOL ONLY	18 (24,0)	17 (24,3)	17 (24,3)
Ct/NCt	*2/16*	*4/13**	*6/11*
SECONDARY SCHOOL NO MATRIC	18 (24,0)	17 (24,3)	17 (24,3)
Ct/NCt	*1/17*	*4/13#*	*4/13 #*
MATRICULATED	22 (29,3)	20 (28,6)	20 (28,6)
Ct/NCtt	*2/20*	*12/8*	*13/7*
TERTIARY	4 (5,3)	3 (4,3)	3 (4,3)
Ct/NCt	*1/3*	*1/2*	*1/2*
TOTAL	75 (100,0)	70 (100,0)	70 (100,0)
Ct/NCt	*6/69*	*22/48*	*25/45*

* - $p < 0,05$; # - $p < 0,001$ I.C. - Intermittent Catheterisation; *Ct/NCt* - Continent/Not Continent

The sex and race of the infant do not appear to play a major role in the achievement of continence, except that more infants from the white group were continent by 5 years of age (Table V.2.51 & V.2.52).

Table V.2.51 CONTINENCE ON I.C. PROGRAMME VS SEX

	INFANT AGE		
SEX	3 YEARS	4 YEARS.	5 YEARS
	n (%)	n (%)	n (%)
MALE	41 (54,7	40 (57,1)	40 (57,1)
Ct/NCt	*1/40*	*12/28*	*14/26*
FEMALE	34 (45,3)	30 (42,9)	30 (42,9)
Ct/NCt	*5/29*	*10/20*	*11/19*
TOTAL	75 (100,0)	70 (100,0)	70 (100,0)
Ct/NCt	*6/69*	*22/48*	*25/45*

I.C. - Intermittent Catheterisation *Ct/NCt* - Continent/Not Continent

Table V.2.52 CONTINENCE ON I.C. PROGRAMME VS RACE

	INFANT AGE		
RACE	3 YEARS	4 YEARS.	5 YEARS
	n (%)	n (%)	n (%)
WHITE	20(26,7)	20(28,6)	20(28,6)
Ct/NCt	*2/18*	*9/11*	*10/10*
MIXED ANCESTRY	43(57,3)	42(60,0)	42(60,0)
Ct/NCt	*4/39*	*12/30*	*14/28*
BLACK	12(16,0)	8(11,4)	8(11,4)
Ct/NCt	*0/12*	*1/7*	*2/6*
TOTAL	75(100,0)	70(100,0)	70(100,0)
Ct/NCt	*6/69*	*22/48*	*25/45*

I.C. - Intermittent Catheterisation *Ct/NCt* - Continent/Not Continent

The status of the upper urinary tract was dependant on the type of neurogenic bladder present. A spastic bladder was the more common finding. A spastic neurogenic bladder was commonly associated with a dilated upper urinary tract (Table V.2.53)

Table V.2.53 UPPER URINARY TRACT STATUS VS BLADDER TYPE

	UPPER URINARY TRACT				
BLADDER TYPE	1 YR	2 YR	3 YR	4 YR	5 YR
	n (%)#	n (%)#	n (%)*	n (%)*	n (%)*
SPASTIC	51 (57,3)	51 (57,3)	51 (60,7)	49 (61,2)	49 (61,2)
UD/D	*23/28*	*23/28*	*23/28*	*12/28*	*21/28*
FLACCID	16 (18,0)	16 (18,0)	12 (14,3)	11 (13,8)	11 (13,8)
UD/D	*15/1*	*15/1*	*11/1*	*10/1*	*10/1*
MIXED	22 (24,7)	22 (24,7)	21 (25,0)	20 (25,0)	20 (25,0)
UD/D	*20/2*	*20/2*	*19/2*	*18/2*	*18/2*
TOTAL	89 (100)	89 (100)	84(100)	80(100)	80(100)
UD/D	*58 /31*	*58/31*	*53/31*	*49/31*	*49/31*

- $p < 0,001$; * - $p < 0,01$ *UD/D* - Undilated/Dilated

as was the need for urological surgery, mainly vesicostomies (Table V.2.54).

Table V.2.54 UROLOGICAL SURGERY VS BLADDER TYPE

	INFANT AGE				
BLADDER TYPE	1 YR	2 YR	3 YR	4 YR	5 YR
	n (%) #	n (%) #	n (%) #	n (%) *	n (%) *
SPASTIC	51(57,3)	51(57,3)	51(60,7)	49 (61,2)	49 (61,2)
R./NR	*19/32*	*19/32*	*19/32*	*18/31*	*18/31*
FLACCID	16(18,0)	16(18,0)	12(14,3)	11(13,8)	11(13,8)
R./NR	*0/16*	*0/16*	*0/12*	*0/11*	*0/11*
MIXED	22(24,7)	22(24,7)	21(25,0)	20925,0)	20(25,0)
R./NR	*0/22*	*0/22*	*0/21*	*0/20*	*0/20*
TOTAL	89(100)	89(100)	84(100)	80(100)	80(100)
R./NR	*19/70*	*19/70*	*19/65*	*18/61*	*18/61*

- $p < 0,01$; * - $p < 0,05$ *R/NR* - Required/Not Required

The type of neurogenic bladder did not appear to influence continence, except that fewer infants with a flaccid bladder attained continence using the intermittent catheterisation programme (Tables V.2.55, V.2.56 & V.2.57).

Table V.2.55 CONTINENCE AT 3 YEARS VS BLADDER TYPE

	URINARY CONTINENCE			
BLADDER TYPE	CONTINENT NO ASSIST.	CONTINENT ON I.C.	NOT CONTINENT	TOTAL
	n (%)	n (%)	n (%)	n (%)
SPASTIC	1(11,1)	4(66,6)	46(66,7)	51(60,7)*
FLACCID	2(22,2)	1(16,7)	9(13,0)	12(14,3)*
MIXED	6(66,7)	1(16,7)	14(20,3)	21(25,0)*
TOTAL	9(100,0)	6(100,0)	69(100,0)	84(100,0)

-$p < 0,05$; I.C. - Intermittent Catheterisation

Table V.2.56 CONTINENCE AT 4 YEARS VS BLADDER TYPE

	URINARY CONTINENCE			
BLADDER TYPE	CONTINENT NO ASSIST.	CONTINENT ON I.C.	NOT CONTINENT	TOTAL
	n (%)	n (%)	n (%)	n (%)
SPASTIC	2(20,0)	15(68,2)	32(66,6)	49(61,3)*
FLACCID	2(20,0)	2(9,1)	7(14,6)	11(13,7)*
MIXED	6(60,0)	5(22,7)	9(18,8)	20(25,0)*
TOTAL	10(100,0)	22(100,0)	48(100,00)	80(100,0)

-$p < 0,05$; I.C. - Intermittent Catheterisation

Table V.2.57 CONTINENCE AT 5 YEARS VS BLADDER TYPE

BLADDER TYPE	URINARY CONTINENCE			
	CONTINENT NO ASSIST.	CONTINENT ON I.C.	NOT CONTINENT	TOTAL
	n (%)	n (%)	n (%)	n (%)
SPASTIC	2(20,0)	16(64,0)	31(68,8)	49(61,3)*
FLACCID	2(20,0)	2(8,0)	7(15,6)	11(13,7)*
MIXED	6(60,0)	7(28,0)	7(15,6)	20(25,0)*
TOTAL	10(100,0)	25(100,0)	45(100,00	80(100,0)

-p < 0,05; I.C. - Intermittent Catheterisation

The LR program reports a linear model for the log odds terms ln(p/1-p). For the categorical explanatory variables of the study the corresponding linear model terms represent relative risks (RR), namely the difference in the log odds ratios associated with moving from one category to another. Confidence intervals that exclude 1 are evidence for significant changes in risk associated with the explanatory variable.

Table V.2.58 indicates under ambulatory status that non-ambulancy or wheelchair-bound conditions are estimated to have 5 times higher risk amongst births originating from areas outside of Cape Town as compared to the Cape Town area. The 95% confidence interval is estimated as (1,40 - 17,48). Similarly, risk of wheelchair boundedness is approximately 55 times greater amongst those with a sensori-motor level in the thoracic and lumbar region above L2 as compared with those with sites below L2.

Table V.2.58 STATISTICS

RESPONSE VARIABLE	EXPLANATORY VARIABLE	RELATIVE RISK	95% CONFIDENCE INTERVAL	P -VALUE
Ambulation	Birth Origin	5,00	1,40 - 17,8	0,0255
	Sensori-Motor level	54,5	5,45 - 544	0,0025
Continence	Social Class II	14,2	2,22 - 91,5	0,0067
	Social Class III	3,79	1,22 - 11,8	
	Birth Origin	4,74	1,47 - 15,2	0,0100
	Delivery Mode	4,10	1,24 - 13,6	0,0221
Griffiths General Quotient	Race Mixed Ancestry	0,286	0,076 - 1,08	0,0505
	Black	0,084	0,012 - 0,583	
	Closure Delay	0,328	0,116 - 0,934	0,0465
	Delivery Mode	3,72	1,27 - 10,9	0,0230

Failure to attain continence has substantially increased risk amongst whites as opposed to those of mixed ancestry and black persons, among rural as opposed to urban origin, and amongst NVD births as opposed to others.

Lower modified Griffiths scores categorisation (<70) has increased risk amongst mixed ancestry and black births than white births amongst those births with a delay to closure greater than 48 hours, and amongst NVD births as opposed to others.

VI DISCUSSION

VI.1 PREVALENCE

In the Cape Town area almost all deliveries occur in a hospital (private or state) or at a midwife obstetric unit, served by the Peninsula Maternal and Neonatal Service (PMNS). The PMNS is a University of Cape Town based urban community perinatal program serving the major section of the community. All births and deaths are routinely registered with the Department of Home Affairs. All cases of neural tube defects delivered in the private or state sector are referred to the Genetic Counselling Service offered by the Department of Human Genetics at the University of Cape Town and to the neurosurgical units either at Red Cross War Memorial Children's or Groote Schuur Hospitals, if surgery is required, and all survivors are followed at the Spinal Defect's Clinic at the Red Cross War Memorial Children's Hospital. In view of the meticulousness of the search and cross-validation between hospitals, services, and departments, I am confident of the completeness of the ascertainment of the neural tube defects.

This study comprised 516 252 births. By comparison, previous studies from Africa have comprised smaller numbers of births. Horner and Lanzkowsky (1966) included 6 502 births and Cornell et al. (1983) 116 859 births in their studies from Cape Town. Singer et al. (1978) reported on 56 110 births from the Cape Province, and from elsewhere in South Africa Grace et al. (1981) included 2 069 births, Kromberg and Jenkins (1982) 29 366 births, and Ncayiyana (1986) 9 142 births. Khan (1965) in his study from Kenya included 3 016 births, Simpkiss and Lowe (1961) from Uganda 2 068 births, and Harrison et al. (1985) from Nigeria 23 512 births.

The overall prevalence of neural tube defects in Cape Town during the study period ranged from a low of 0,63/1 000 births in 1992 to a high of 1,74/1 000 births in 1985. These are similar to figures from first world countries (Elwood, 1973; Janerich,

1973) and figures from other countries in Africa (Khan, 1965; Harrison et al., 1985) and also to those previously reported locally (Horner and Lanzkowsky, 1966; Singer et al., 1978; Kromberg and Jenkins, 1982; Cornell et al., 1983). However, they are low in comparison to the data from an area in the Transkei (Ncayiyana, 1986).

The prevalences for the various racial groups in the Cape Town area are significantly different, being highest for the whites and lowest for blacks. The overall rate for whites, while similar to figures quoted for the British Isles (EUROCAT, 1991), is lower than that previously reported for Cape Town (Cornell et al., 1983). However the data of Cornell et al. (1983) only included whites attending state hospitals, to which affected cases may well have been referred for delivery in a state hospital, thus skewing the data.

The prevalence for blacks is higher than that previously found in Cape Town by Cornell et al. (1983), but similar to that reported for the Cape Province by Singer et al. (1978), and elsewhere in South Africa (Grace et al., 1981; Kromberg and Jenkins, 1982), Kenya (Khan, 1965), Uganda (Simkiss and Lowe, 1961), Nigeria (Harrison et al., 1985), and for African Americans (Myrianthopoulos and Melnick, 1987), but lower than the figure reported from an area in the Transkei (Ncayiyana, 1986). This high prevalence from the Transkei may represent a geographical or genetic trend in that population, when one considers that the Black inhabitants of Cape Town are mostly migrants from the Transkei and Ciskei.

The neural tube defect prevalence for those of mixed ancestry is similar to data previously reported for this region (Horner and Lanzkowsky, 1966; Singer et al., 1978; Cornell et al., 1983) and has remained relatively constant for the study period. The frequency is however, lower than that reported by Grace et al. (1981) from Durban, where the size of the population of mixed ancestry is far smaller than that in Cape Town and their genetic origins are somewhat different from those in the Cape. The Natal mixed ancestry population is derived from Afro/European/Indian sources, whereas those from the Cape have European/Khoi-Khoi/Malay origins.

The overall prevalence of all forms of NTDs was highest for the whites and lowest for the blacks, with the exception of the overall prevalence of myelomeningocele where the lowest was in the mixed ancestry group. This trend is similar to data from the Birth Defects Monitoring Program in the United States (Khoury et al. 1982).

Overall the rate for anencephaly for whites is lower, but that for spina bifida is similar to previous reports for this region (Horner and Lanzkowsky, 1966; Singer et al., 1978; Cornell et al., 1983). In the mixed ancestry group the overall rate for anencephaly is similar to that of Singer et al. (1978) and Cornell et al. (1983), but lower than that found by Horner and Lanzkowsky (1966). For spina bifida, however it is slightly higher than that previously reported (Horner and Lanzkowsky, 1966; Singer et al., 1978; Cornell et al., 1983). The rate for anencephaly for blacks during this period is lower and that previously quoted (Singer et al., 1978; Cornell et al., 1983), whereas for spina bifida the rate is similar to that of Singer et al.(1978), but higher than that reported by Cornell et al. (1983). Both these figures are lower than those from the Transkei (Ncayiyana, 1986).

The ratio for spina bifida:anencephaly was highest for blacks and lowest for those of mixed ancestry. The ratio for blacks is higher than previously reported from Cape Town (Cornell et al., 1983), the Cape Province (Singer et al., 1978) or the Transkei (Ncayiyana, 1986), and higher than that reported for the African American (Khoury et al., 1982). The ratio for those of mixed ancestry is higher than that reported from Cape Town by Horner and Lanzkowsky (1966) or Cornell et al. (1983) but lower than that reported by Singer et al. (1978) for the Cape Province. The ratio for whites is the highest reported from Cape Town (Horner and Lanzkowsky, 1966; Cornell et al., 1983), the Cape Province (Singer et al., 1978) or Johannesburg (Kromberg and Jenkins, 1982), and higher than that reported from first world countries (Janerich, 1973; Elwood, 1973; Khoury et al. 1982).

The female predominance for spina bifida, anencephaly and total NTD is similar to that reported from first world countries (Janerich, 1975; Khoury et al., 1982; Windham and

Edmonds, 1982; Dolk et al., 1991). This predominance has also been found in the Transkei (Ncayiyana, 1986).

The association of an increasing prevalence of neural tube defects with increasing social class has previously been documented, suggesting a possible nutritional or infective aetiology (Record and McKeown, 1949; Coffey and Jessop, 1957; Fedrick, 1970). In Cape Town the highest prevalence is in the white population group, the majority of whom are members of the upper socio-economic classes with some British ancestry, so effects of nutrition and infection are confounded with genetic factors, which may be more important.

During the twenty year period under review, no trend could be demonstrated in neural tube defect prevalence unlike the decreases ascertained in the United States (Janerich, 1973; Windham and Edmonds, 1982; Snyder et al., 1991), and in Britain (Elwood, 1973; Lorber and Ward, 1985). However, more recently, there are suggestions that an upward trend may be evident in certain cities in the United Kingdom (EUROCAT, 1991) and in Hungary (Czeizel, 1983).

The relationship to maternal age is well demonstrated in this study. There is a higher prevalence at both ends of the maternal age range. This pattern is in keeping with a U-shape described by Janerich in 1972 for American women. The higher prevalence in older women has also been documented in Britain (Bound et al., 1991).

A seasonal prevalence is well shown locally and is similar to that previously reported by McKeown and Record (1951) and Leck (1974) and suggested by local data from Singer et al. (1978). It is interesting that the increase in conceptions of neural tube defects occurs during similar periods both in the northern and southern hemispheres albeit during different seasons. The increased prevalence of neural tube defects seen in the first born and higher birth order is similar to that shown in studies from first world countries (Fedrick, 1970).

Eight percent of the NTD pregnancies locally were legally terminated. This figure is far lower than that reported from first world countries, where up to 75% are terminated, more so in cases of anencephaly than spina bifida where the antenatal diagnosis is made earlier and more frequently (Ferguson-Smith, 1983; EUROCAT, 1991). Until mid-1981 maternal alpha-fetoprotein and sonography were used in the antenatal diagnosis of NTDs, but since then only sonography has been employed. The actual effect of the prenatal screening programme has been questioned by some (Shepherd, 1983), suggesting that the declining trend for neural tube defects seen in first world countries actually preceded their antenatal screening programmes. A problem locally is that the majority of our mothers book after 20 weeks gestation when the alpha-fetoprotein screen is not as reliable or often the prenatal diagnosis is made once the foetus is viable and termination a problem (Van Coevereden De Groot, 1995). The inception of the Fetal Abnormality Group in 1983 coincides with the increase in our prevalence of NTDs, but as this rise occurred in our spina bifidas and not the occult NTDs (anencephalics), it is suggestive that the increase in surveillance was not responsible for the rise.

This study is the first comprehensive analysis of neural tube defect prevalence over a twenty year period in Africa. No trends could be demonstrated for all neural tube defects but there are considerable racial differences. Parity, maternal age, and sex ratios exhibit patterns similar to those seen in most first world countries. A seasonal variation has been shown which apparently differs from that reported for the Northern Hemisphere.

VI.2 OUTCOME

The paediatric neurosurgical service at Groote Schuur and Red Cross War Memorial Children's Hospitals is the only service of this nature available in the Cape Province and for many hundreds of kilometres outside of Cape Town. Although neurosurgical services do exist in state and private hospitals in other major centres outside of Cape Town (Kimberley, Port Elizabeth and East London), they are not specialised in paediatric

neurosurgery. Occasionally, closures of myelomeningoceles do take place in these centres, but the majority of newborns with a congenital myelomeningocele are referred to Cape Town for closure. On occasions this referral extends beyond the borders of the Cape Province to neighbouring provinces and countries. What preselection of patients took place in these centres or in rural hospitals outside of Cape Town is unknown, and for this reason only the outcome of patients who underwent surgery in Cape Town has been studied.

More patients came from outside of Cape Town (65 vs. 53) and those with higher lesions tended to be referred from these areas, which would suggest that less preselection took place than was expected. More of the referred patients were black, with parents of a lower level of education and from social class III. This would be expected as the majority of rural patients are farm labourers. The majority of the black patients in this group came from the Eastern Cape areas of the Ciskei and Transkei. The association of myelomeningocele with decreasing social class had been previously well documented (Record and McKeown, 1949; Coffey and Jessop, 1957; Fedrick, 1970). This also pertained to the prevalence section of this study.

As expected the majority of the mothers in both groups were in their twenties, except that a higher number of teenage pregnancies occurred in the Cape Town social class III group, which may reflect the effects of urbanisation.

The educational profile of the parents is in keeping with their social class, with the majority of these receiving no formal education coming from outside of Cape Town and belonging to social class III. This is a reflection of the availability of education to the rural community. Maternal and paternal education profiles are very similar.

An antenatal diagnosis of a myelomeningocele was made in only four patients who required surgery. In the early part of this study serum alpha-fetoprotein screening was being done in the area served by the Peninsula Maternal and Neonatal Service of the

University of Cape Town. No local infants operated on were detected in the screen and this suggests that those affected pregnancies detected may have been terminated. Alternatively, because most of the local mothers attend antenatal clinic for the first time well into their second trimester, the interpretation of the screen may have been difficult. The two cases with a family history of a previous neural tube defect were from outside of the Cape Town area and were not screened antenatally. The majority of the mothers (80%) were not screened for a neural tube defect. The high success rate (95%) using antenatal ultrasound screening for neural tube defects suggested by studies from first world countries (Nicolaides et al., 1986; Campbell et al., 1987; Penso et al., 1987; Thiagarejah et al., 1990), is not seen locally. A possible explanation is that infants diagnosed as having a NTD were terminated, but in the prevalence study, over the twenty year period only 34 (10%) legal terminations were carried out for myelomeningocele, and the majority of these were after 1983 with the inception of the Fetal Abnormality Group at Groote Schuur Hospital. A more likely explanation is that the inexperience of the ultrasonographer and the equipment used during the early part of the study resulted in the diagnoses not being made. The number of affected cases was higher in first born infants, in keeping with reports from the United Kingdom (Fedrick, 1970) and New Zealand (Borman and Cryer, 1993), but not those from Israel (Naggan, 1971).

The majority of the affected infants were delivered under medical supervision. Indications for caesarean section did not appear to be related to a large head circumference at birth, with only 40% of those with a head circumference over the 90th centile (Lubchenko) requiring delivery by caesarean section. However, those delivered by caesarean section had a better chance of being continent and having a modified Griffiths general quotient >70, in keeping with the findings of Bensen et al. (1988). Delivery by caesarean section was unrelated to successful ambulation, as was found by Cochrane et al. (1991), but not by Shurtleff et al. (1987).

Although 16 infants were delivered preterm, only 2 were < 32 weeks gestation. Both of them died from complications related to their prematurity (hyaline membrane disease and

necrotizing enterocolitis). Fifteen (12,7%) were growth retarded, but this did not appear to affect their outcome.

There was some correlation between the anatomical site of the myelomeningocele and the sensory motor level at birth. The majority of the infants had lumbar or sacral sensori-motor levels, which is similar to that reported by McLone (1981) and others. Mid-lumbar (L3 & L4) and lower lumbar (L5) lesions were more likely to come from outside of the Cape Town area. The only cervical myelomeningocele had a thoracic sensori-motor level and required an Arnold Chiari decompression. The infant died of recurrent apnoea.

The more common congenital anomalies included congenital talipes equinovarus (CTEV), strabismus, congenital dislocation of the hip (CDH) and kyphoscoliosis. There appeared to be a relationship between the sensori-motor level and CTEV and CDH, which was significant only for CTEV, being more common with a lumbar sensori-motor level. CDH was more commonly associated with a mid-lumbar sensori-motor level (L3 & L4), a finding previously reported by Carroll and Sharrard (1972). Kyphoscoliosis only occurred in patients with mid-lumbar, lower lumbar and upper sacral lesions, which is contrary to the increased risk reported by Samuelsson and Eklof (1988) in infants with a sensori-motor level above L3.

In contrast to some first world studies, only one third of the patients had their lesion closed within 24 hours of birth (cf. McLone et al., 1981, 91% closed within 24 hours). Within 48 hours of birth the figure improved to 50%. Significantly, the majority came from the Cape Town area. Delays in the referral of country patients were mostly transport related, and those referred much later were probably survivors of initial conservative management at other hospitals.

Seventy seven percent of infants had hydrocephalus requiring some form of drainage procedure (mainly ventriculo-peritoneal shunts), which is similar to the findings of others

(Stein et al., 1974; McLone et al ., 1981; Soare and Raimondi, 1977). Fifty (42,3%) required only a single ventriculo-peritoneal shunt and 29 (24,6%) needed revisions of the procedure. The presence of hydrocephalus requiring shunting was more commonly associated with a sensori-motor level in the lumbar and thoracic regions and a birth head circumference >90th centile (Lubchenko), a finding previously reported by Laurence (1964).

Only 20% of the ventriculo-peritoneal shunts were placed within 7 days of birth, the rest being placed within the next 7 months of life. Shunt failure, requiring revision occurred most commonly in the first year of life similar to the findings of Liptak et al. (1985).

At five years of age three quarters of the children had Modified General Quotients >70 on the Griffiths Scales of Mental Development, although 15% were in the borderline range (70 - 79). Of those with scores below 70, four fifths were mildly handicapped (50 - 69) and one fifth were moderately handicapped (30 - 49). The scores on the locomotor sub-test were lower than the other sub-tests. Personal/social and speech/hearing were marginally higher than eye-hand, performance and practical reasoning. Most children with myelomeningocele have selective cognitive disabilities and score better on verbal than performance scales (Liptak et al., 1988).

Hydrocephalus has been recognised as a limiting factor in the intellectual functioning of children with myelomeningocele. A number of studies have shown intellect in non-shunted children to be normal and well above those requiring shunting (Badell-Ribera et al., 1966; Spain, 1974; Soare and Raimondi, 1977; McLone et al., 1982; Mapstone et al., 1984). Our findings agree with this, although our mean developmental quotient for non-shunted patients of 80 is lower than those of Mapstone et al. (1984), McLone et al. 1982) and Spain (1974). In our study those who required only one shunt were no different to those not shunted, whereas the group shunted more than once functioned significantly lower in all sub-tests. There is clearly a relationship between central nervous system infection and developmental performance, a finding previously reported by Hunt

and Holmes (1976), McLone et al. (1982), and Casari and Fantino (1992), but not reported by Charney et al. (1985) and Sutton et al. (1986). Much of the developmental fall off is therefore due to infection as a complication of shunting, in addition to the adverse effects of shunt blockage.

Serial testing was carried out from infancy to five years of age. A developmental quotient was obtained using the DEI at one, two, three and four years. At five years the children were administered the Griffiths Scales of Mental Development. A high correlation, which improved over the years, was found between the DEI and the Griffiths Scales. This finding agrees with that of Fishman and Palkes (1974) who demonstrated a correlation between the Cattel Infant Scales at 18 months and the Stanford-Binet at five years. Although the DEI was developed to plan and direct early intervention in this group of patients, it appears to have predictive value for later intelligence which is helpful in parent counselling.

The overall ambulation rate in this study was 71,3%. Similar ambulation rates have been reported from selected series (Stark and Drummond, 1973; Lorber and Salfield, 1981) and from unselected series (McLone et al., 1985; Beks et al., 1966; Ames and Schut, 1972). The sensori-motor level of the lesion is an important factor affecting ambulation. This has previously been shown by others (Hoffer et al., 1973; Asher and Olson, 1983). A major factor in the mid-lumbar (L3 and L4) lesions is hip stability and this is reflected in the delay in ambulation and the increased requirement for orthopaedic surgery in this group. This has also been reported elsewhere by Carroll and Sharrard (1972), Hoffer et al. (1973), Asher and Olson (1983), and locally by Fraser et al. (1992). Children from the Cape Town area are ambulant earlier, which may be because of delays in orthopaedic management or the maintenance of appliances used for ambulation in the group from outside of Cape Town, who do have a higher number of mid-lumbar and lower lumbar lesions.

Intelligence appeared to play a significant role in ambulation in this study, which may be a reflection of the extent of their overall handicap, a factor previously found not to affect ambulation in mid-lumbar lesions (Fraser et al., 1992).

Thirty five (44%) of the infants were continent at five years of age, 25 on an intermittent catheterization programme and 10 without assistance. Purcell and Gregory (1984) reported a lower success rate (24%) and Uehling et al.(1985) a higher success rate (81%), but their patients had been on the programme for a minimum of 5 years. The sensori-motor level, level of intelligence, birth origin and social class all played a role in urinary continence. Lapides et al. (1972) previously suggested that intelligence influenced the success of intermittent catheterization. Those living within the Cape Town area were more successful in becoming continent on the intermittent catheterization programme because of the availability of medical and nursing expertise when problems arose on the programme. They were also more readily available for initial investigations and commencement on the programme. This trend is also evident for social class and maternal education, reflecting that most families from outside of Cape Town are farm labourers, of social class III and only one third of whom have more than primary school education.

Thirty eight percent of the children had a dilated upper urinary tract which is higher than that reported by Eckstein (1965). A dilated upper urinary tract is more commonly associated with a spastic neurogenic bladder (increased urethral sphincter resistance). To preserve the upper urinary tract, the more common surgical procedure is a suprapubic vesicostomy and non-surgical drainage intermittent urethral catheterization. In the case of the spastic bladder where urethral sphincter resistance is elevated, more success would be expected in attaining continence on the intermittent catheterization programme. However, the type of bladder did not seem to play a major role in the attainment of continence in these patients.

This is the first comprehensive study on the outcome of children with myelomeningocele over a five year period in Africa. Most previous studies from first world countries have used a variable follow-up period to assess outcome, in particular intelligence, therefore relying on a number of different developmental and intelligence tests. This makes data interpretation and comparison difficult. All children followed-up in this study were formally assessed at five years of age using the Griffiths Scales of Mental Development. Factors that played an important role in the outcome of these children were the delay in neurosurgery, central nervous system infection, birth origin, social class and maternal education.

VII MANAGEMENT OF NEURAL TUBE DEFECTS IN THE WESTERN CAPE

VII.1 PRINCIPLES

Management must be based on the Primary Health Care approach which enhances -

- equity;
- comprehensiveness - a fully integrated (promotive, preventive, curative and rehabilitative) multidisciplinary approach;
- community participation and empowerment;
- affordability and sustainability.

(Declaration of Alma Ata, 1978)

The aims are to limit disability, to achieve optimal development and social integration of the child.

VII.2 PRACTICE

VII.2.1 PREVENTION

Prevention of neural tube defects and their recurrence has been taken a major step forward with the discovery that pre- and periconceptional supplementary folate can prevent their occurrence and recurrence. It is now recommended that all women of childbearing age who are capable of becoming pregnant should receive folate supplementation to reduce the risk of having a neural tube defect affected pregnancy. One of the major problems in instituting the recommendation in the Western Cape, is that most mothers from the lower socio-economic group first attend antenatal clinic well into their second trimester of pregnancy. This could be overcome by the fortification with folate of certain staple foods such as bread flour or rice (Seller, 1994).

Ideally antenatal screening for neural tube defects, including alpha-fetoprotein, acetyl cholinesterase and fetal sonography should be freely available. As mentioned one of the problems is the mother first attending antenatal clinic well into her second trimester when screening using alpha-fetoprotein is difficult to interpret, and if the diagnosis of a NTD affected pregnancy be made, a legal termination of the pregnancy, should she request it, may no longer be possible or safe for the mother. Genetic counselling should be and is available to any mother who has a family history of a neural tube defect or has or has had a neural tube defect affected pregnancy. As the most frequently used method of antenatal diagnosis for an NTD is antenatal sonography, it should be offered as a screening method for NTDs. Equipment and training of the personnel involved in antenatal sonographic programmes in peripheral centres in the antenatal diagnosis of NTDs should be reviewed to improve the diagnostic yield.

Education of future mothers, especially at schools, regarding the use of family planning, folate supplementation, the importance of early attendance at antenatal clinic and the risks of teenage pregnancy, is an important and often neglected method of prevention.

No discussion on prevention of NTDs would be complete without some reference to ethical issues. Selective abortion and non-treatment of infants with spina bifida may be regarded as secondary preventive measures. Although these measures are legally sanctioned in South Africa, questions arise which stem from ethical, moral and religious beliefs. The decisions as to whether to proceed with these preventive measures must rest with the various religious groups and indeed with the families themselves.

VII.2.2 DELIVERY

Deliveries should be conducted under medical supervision in a hospital or obstetric facility. When the antenatal diagnosis of a myelomeningocele has been made, the role of a caesarean section as a method of delivery is a consideration, especially if the

presentation is breech or if the head size in the presence of hydrocephalus is too large for a safe vaginal delivery.

VII.2.3 EARLY MANAGEMENT

Optimally, when the antenatal diagnosis had been made, transfer of the mother from a rural or peripheral area to a larger centre where neurosurgical expertise is be available for the infant, should take place. Once delivered, the infant with a myelomeningocele needs referral to neurosurgical expertise within the first twenty four hours. While awaiting transfer the lesion should be kept clean and moist, consent for possible surgery obtained, and the parents counselled appropriately.

Closure of the lesion should be carried out in a paediatric neurosurgical unit if possible. In the event of any delay in transfer or closure of the lesion the use of prophylactic antibiotics should be considered. Post surgery monitoring for ensuing hydrocephalus must be carried out and a ventriculo-peritoneal shunt inserted if required.

VII.2.4 MULTIDISCIPLINARY FOLLOW-UP

Follow-up of a child with a myelomeningocele requires a multidisciplinary team, which should preferably be available at the same venue on the same day, saving on transport expenses and time. Neurosurgical expertise should be available for the long term management of ventriculoperitoneal shunt problems and other neurosurgical problems encountered in these children. Orthopaedic input is necessary for the ongoing management of the orthopaedic problems experienced by these children and in association with an orthoptist monitoring and modifying of their orthopaedic appliances. Urological expertise is required for the long term management of their neurogenic bladder and incontinence, and monitoring of their upper urinary tract. Paediatric involvement in the

form of developmental guidance should be available for ongoing evaluation of developmental problems should they arise. A social worker should be available for counselling of families when necessary.

VII.2.5 DEVELOPMENTAL ASSESSMENT / INTERVENTION

The use of the DEI screening assessment has proved useful and results correlate well with the more formal Griffiths Scales of Mental Development. It is quick and easy to use and can therefore be administered frequently to follow any specific delay in development that has been detected. Once a delay has been detected, specific referral may be indicated e.g. audiology/speech therapy if a speech and hearing delay is found, or referral to a toy library facility if only a fine motor adaptation delay is found. Usually the delay if present is of a more global nature in children with myelomeningocele and this is often associated with over protection by parents and emotional immaturity. Early pre-school attendance is helpful in this situation. Unfortunately most pre-schools in Cape Town do not accept children who are not toilet trained making it difficult for the child with myelomeningocele, as the majority at this age are still incontinent of urine. Discussing the specific case with the school often helps but is not always successful. Special schools for physically handicapped children with pre-school facilities are available and have been accommodating in the past for children with myelomeningocele, but their facilities are limited. Community based rehabilitation (CBR) is an important form of intervention particularly in areas where formal services are limited. The focus is on community involvement and the use of simplified rehabilitation technology. It is carried out by appropriately trained community rehabilitation workers who are often family members (Helander, 1984).

VII.2.6 SCHOOLING

To date most schools in the Western Cape do not accommodate physically handicapped or incontinent children. The majority of children with myelomeningocele in this area have attended special schools for physically disabled children on condition they are capable of a normal education program. As mentioned the facilities of this nature are only available in large centres and are limited, especially hostel facilities. Children, in particular those living some distance from a large centre, are often denied education because of lack of space at a special school, and schools in their own area refuse them admission because they too are overcrowded and because they don't have a school nurse on the premises to cope with the handicapped child or with urinary incontinence. The importance of inclusive education has been recognised (Salamanca Document, 1994). Inclusive (mainstream) schools recognise and respond to the diverse needs of their pupils, accommodating both different styles and rates of learning and ensuring quality education to all through appropriate curricula, organisational strategies and partnerships with their communities. It plays a role in minimising handicap and allowing the children to be productive adults. It is unfortunate that mainstreaming is not usual in schools in South Africa as most schools do not have a school nurse, lack facilities for the handicapped, and not all schools have remedial or special educational facilities available on the premises.

VII.2.7 COUNSELLING

Counselling of a family with a child with an NTD should begin as soon as the diagnosis is known. Antenatal counselling is usually in the form of genetic counselling with the possibility of a termination of pregnancy. Often cases of myelomeningocele are only diagnosed at delivery and the initial counselling starts in the post delivery period when consent for surgical correction of the defect is obtained. The long term counselling of the family should be conducted by a social worker experienced in this field, who should be

available to the family to discuss the expected outcome, with the backup of specialist medical expertise (neurosurgical, urological, orthopaedic, paediatric).

The adolescent with myelomeningocele presents other problems for which counselling is needed. Compounding the usual complex changes of adolescence and sexuality, most have a poor self-image, are often emotionally immature and some may even have below average intelligence, requiring skilled counselling.

VII.2.8 SUPPORT GROUP

The establishment of a support group for families with a child with myelomeningocele is helpful. Sharing and the realisation that there are other children with similar problems is beneficial in the long-term. The group should be structured and can be used as a venue to educate parents and children and as a forum to discuss problems in their long-term management. Social events should also be organised to encourage attendance and communication between families.

The use of a group newsletter as a form of communication to inform parents of meetings, availability and obtainability of appliances and changes to other management protocols, to act as a mouthpiece for both families and the medical support teams, and to maintain contact with families living a long distance away who are unable to attend meeting regularly, should be encouraged. Smaller groups in areas where a number of families live, especially in areas where families may encounter difficulties attending meetings because of distance or lack of transport or finance, could be established to maintain communication.

The group, if representative of the myelomeningocele community could be used to lobby at local government level to help improve facilities, services, schooling and employment opportunities for the physically disabled.

VII.3 SERVICES FOR THE WESTERN CAPE

VII.3.1 TERTIARY CARE

These facilities exist only exist in the Cape Peninsula area at Groote Schuur, Tygerberg and the Red Cross War Memorial Children's Hospitals. The majority of myelomeningoceles are closed by the Paediatric Neurosurgery Department at the Red Cross War Memorial Children's Hospital, although some are closed at the other hospitals. Urological and orthopaedic investigation and management of the individual with a myelomeningocele are conducted at all three hospitals, but only paediatric patients are seen at the Red Cross War Memorial Children's Hospital. The only multidisciplinary clinic for spinal defects is run at the Red Cross War Memorial Children's Hospital and follows the majority of paediatric patients.

VII.3.2 SECONDARY CARE

Secondary care is available at all regional, district and day hospitals in the Western Cape and access to these hospitals is available to all patients. Regional hospitals have specialists who are able to attend to most management problems in myelomeningocele. In areas where there are sufficient numbers of children with the condition the development of a multidisciplinary clinic should be encouraged. Specialist medical expertise available either in the regional or district hospital or from private practice should be utilized, and if non-specialists from these hospitals are interested, they could be trained at a larger centre to run the service.

Orthopaedic surgery has an ongoing service to regional hospitals in peripheral areas, with regular visits by either medical or paramedical personnel who attend to problems with orthopaedic appliances.

An area of concern is the rate of incontinence in these children by 5 years of age, which makes school admission difficult. Education of parents and children are crucial for success in this area. This could be achieved using nursing staff who are adequately trained in this area, who could educate parents and the children, and be available for any problems with the intermittent clean catheterization programme. The nursing staff concerned could be trained in tertiary care facilities, be based at local/regional/clinics/hospitals, and be updated by visits of clinic personnel from tertiary facilities.

VII.3.3 PRIMARY CARE

Primary care is provided at the local clinics in the Western Cape. It is in the process of being improved. These clinics are primarily staffed by nurses. The prevention of NTDs as part of a primary health care programme is important. Making the population aware of the risk factors, the benefits of using family planning, the use of folate in the prevention of the occurrence and recurrence of NTDs, attending antenatal clinic early in a pregnancy are all important. This could either be done at a primary care clinic, through the media or more importantly at schools.

The rapid developmental screening test (DEI) used by the Developmental Clinic and Spinal Defect's Clinic at the Red Cross War Memorial Children's Hospital has been shown to be reliable in predicting the intellectual abilities of infants with myelomeningocele and is therefore useful in the placement of the children for their educational needs. The test is simple and could be used in a multidisciplinary clinic or at a community based clinic by medical, paramedical or nursing staff.

VII.4 CONCLUSIONS

The management of NTD in the Western Cape Province, with a population of over four million, will require co-operation between the Departments of Health, Education and Social Services. The Provisional Health Plan (1995) on health expressed a commitment to primary health care. Services will be accessible, comprehensive and based on equity. As far as NTD are concerned, emphasis is needed on family planning, maternal education, folate supplementation and antenatal care. Good perinatal care and optimal early management of all cases is essential. Ongoing management will require the co-ordinated use of primary, secondary and tertiary services as outlined above. In addition, community based rehabilitation should be developed as part of the overall strategy. It should be seen as a specific approach within the community, aimed at rehabilitation and social integration of children with myelomeningocele.

Education should be based on the guiding principle that all children must be accommodated regardless of their physical, intellectual or social conditions. Where possible children should attend their neighbourhood schools with appropriate support. Placement in special schools is considered on a case-by-case basis. Special schools can also serve as local resource centres extending their expertise to inclusive education (mainstream) schools. All teachers require pre- and inservice training in aspects of NTDs so that the children's needs can be met. Parent partnership is necessary and a positive attitude on the part of parents favours school and social integration. Community involvement should be sought not only for community based rehabilitation but also to supplement in-school activities. Above all public awareness is crucial and the mass media can play a powerful role in promoting positive attitudes towards children with myelomeningocele.

Social services play a role in providing family support and supplementary grants where needed and in co-ordinating voluntary associations and non-governmental organisations. They also encourage and support community programmes. Finally the role of research in

promoting knowledge and awareness of NTDs is recognised. It is hoped that this study will in some measure contribute to the services and to the well being of the children with myelomeningocele.

APPENDIX 1

TABLE A.1 - THE NUMBER OF NTDS BY RACE IN CAPE TOWN, 1973 - 1992.
(ANENCEPHALY AND ENCEPHALOCELE)

YEAR	ANENCEPHALY				ENCEPHALOCELE			
	W	MA	B	T	W	MA	B	T
1973	2	1	-	3	1	1	1	3
1974	4	5	1	10	2	2	1	5
1975	3	5	-	8	-	2	-	2
1976	1	3	3	7	-	-	1	1
1977	3	5	2	10	1	2	-	3
1978	3	4	3	10	-	-	-	-
1979	2	3	1	6	1	-	-	1
1980	2	1	1	4	1	2	-	3
1981	2	2	1	5	-	1	-	1
1982	3	2	1	6	1	-	-	1
1983	2	4	3	9	-	2	1	3
1984	4	11	-	15	-	1	-	1
1985	1	7	4	12	1	-	1	2
1986	-	3	3	6	-	-	-	-
1987	-	8	3	11	1	-	-	1
1988	1	8	3	12	1	3	1	5
1989	-	4	2	6	1	2	-	3
1990	1	8	-	9	-	2	-	2
1991	2	8	-	10	-	1	1	2
1992	-	3	2	5	-	-	2	2

W - White; MA - Mixed Ancestry; B - Black; T - Total;
NTDS - Neural Tube Defects.

TABLE A.2 - THE NUMBER OF NTDS BY RACE IN CAPE TOWN, 1973 - 1992.
(MYELOMENINGOCELE, OTHER FORMS OF SPINA BIFIDA AND TOTAL SPINA BIFIDA)

YEAR	MYELOMENINGOCELE				OTHER				SPINA BIFIDA			
	W	MA	B	A	W	MA	B	A	W	MA	B	A
1973	5	5	1	11	-	3	-	3	5	8	1	14
1974	2	5	3	10	-	-	1	1	2	5	4	11
1975	2	11	4	17	1	3	-	4	3	14	4	21
1976	1	8	1	10	1	2	-	3	2	10	1	13
1977	3	11	3	17	1	1	1	3	4	12	4	20
1978	5	6	1	12	-	1	-	1	5	7	1	13
1979	5	7	1	13	-	3	-	3	5	10	1	16
1980	4	8	1	13	-	2	1	3	4	10	2	16
1981	4	11	1	16	1	3	-	4	5	14	1	20
1982	5	8	4	17	2	-	-	2	7	8	4	19
1983	3	10	7	20	-	2	1	3	3	12	8	23
1984	5	13	7	25	2	2	2	6	7	15	9	31
1985	11	8	10	29	1	5	2	8	12	13	12	37
1986	3	6	5	14	-	2	1	3	3	8	6	17
1987	2	4	4	10	1	-	-	1	3	4	4	11
1988	2	8	5	15	2	2	-	4	4	10	5	19
1989	2	9	14	25	1	2	-	3	3	11	14	28
1990	6	15	9	30	-	2	1	3	6	17	10	33
1991	1	13	10	24	-	1	1	2	1	14	11	26
1992	2	6	3	11	-	1	1	2	2	7	4	13

W - White; MA - Mixed Ancestry; B - Black; T - Total;
NTDS - Neural Tube Defects.

TABLE A.3 - THE TOTAL NUMBER OF NTDS AND BIRTHS BY RACE IN CAPE TOWN 1973 - 1992.

YEAR	TOTAL NTD				TOTAL BIRTH			
	W	MA	B	T	W	MA	B	A
1973	8	10	2	20	3 813	14 583	4 035	22 431
1974	8	12	6	26	3 622	14 029	4 368	22 019
1975	6	21	4	31	3 310	12 778	4 128	20 216
1976	3	13	5	21	3 212	12 545	4 206	19 963
1977	8	19	6	33	2 891	12 326	4 014	19 231
1978	8	11	4	23	2 778	12 569	4 160	19 507
1979	8	13	2	23	2 707	13 165	4 074	19 946
1980	7	13	3	23	2 742	13 841	5 519	22 102
1981	7	17	2	26	2 892	14 983	6 382	24 257
1982	11	10	5	26	2 919	16 253	6 922	26 094
1983	5	18	12	35	2 823	16 644	7 876	27 343
1984	11	27	9	47	2 608	17 340	8 978	28 926
1985	14	20	17	51	2 396	17 291	9 683	29 370
1986	3	11	9	23	2 258	16 699	9 974	28 931
1987	4	12	7	23	2 074	16 767	10 134	28 975
1988	6	21	9	36	2 047	17 268	11 104	30 419
1989	4	17	16	37	1 988	17 129	10 965	30 082
1990	7	27	10	44	1 877	17 065	12 489	31 431
1991	3	23	12	38	1 620	17 860	13 615	33 095
1992	2	10	8	20	1 398	17 371	13 145	31 914

W - White; MA - Mixed Ancestry; B - Black; T - Total;
NTDS - Neural Tube Defects; MOH - Medical Officer of Health;
PMNS - Peninsula Maternal and Neonatal Service;

TABLE A.4 - THE PREVALENCE OF NTDS BY RACE IN CAPE TOWN, 1973 - 1992. (ANENCEPHALY AND ENCEPHALOCELE)

YEAR	ANENCEPHALY				ENCEPHALOCELES			
	W	MA	B	T	W	MA	B	T
1973	0,53	0.07	-	0,13	0,26	0,07	0,25	0,13
1974	1,10	0,36	0,23	0,45	0,55	0,14	0,23	0,23
1975	0,91	0,39	-	0,40	-	0,16	-	0,10
1976	0,31	0,24	0,71	0,35	-	-	0,24	0,05
1977	1,04	0,41	0,50	0,52	0,35	0,16	-	0,16
1978	1,08	0,32	0,72	0,51	-	-	-	-
1979	0,74	0,23	0,25	0,30	0,37	-	-	0,05
1980	0,73	0,07	0,18	0,18	0,37	0,15	-	0,14
1981	0,69	0,13	0,16	0,21	-	0,07	-	0,04
1982	1,03	0,12	0,15	0,23	0,34	-	-	0,04
1983	0,71	0,24	0,38	0,33	-	0,12	0,13	0,11
1984	1,53	0,63	-	0,52	-	0,06	-	0,04
1985	0,42	0,41	0,41	0,41	0,42	-	0,10	0,07
1986	-	0,18	0,30	0,21	-	-	-	-
1987	-	0,48	0,30	0,38	0,48	-	-	0,04
1988	0,49	0,46	0,27	0,40	0,49	0,17	0,09	0,16
1989	-	0,23	0,18	0,20	0,50	0,12	-	0,10
1990	0,53	0,47	-	0,29	-	0,12	-	0,06
1991	1,23	0,45	-	0,30	-	0,06	0,07	0,06
1992	-	0,17	0,15	0,16	-	-	0,15	0,06

W - White; MA - Mixed Ancestry; B - Black; T - Total;
NTDS - Neural Tube Defects

TABLE A.5 - THE PREVALENCE OF NTDS BY RACE IN CAPE TOWN, 1973 - 1992. (MYELOMENINGOCELE, AND OTHER FORMS OF SPINA BIFIDA)

YEAR	MYELOMENINGOCELE				OTHER			
	W	MA	B	T	W	MA	B	T
1973	1,31	0,34	0,25	0,49	-	0,21	-	0,13
1974	0,55	0,36	0,69	0,45	-	-	0,23	0,05
1975	0,60	0,86	0,97	0,84	0,30	0,24	-	0,20
1976	0,31	0,64	0,24	0,50	0,31	0,16	-	0,15
1977	1,04	0,89	0,75	0,88	0,35	0,08	0,25	0,16
1978	1,80	0,48	0,24	0,62	-	0,08	-	0,05
1979	1,84	0,53	0,25	0,65	-	0,23	-	0,15
1980	1,46	0,58	0,18	0,59	-	0,15	0,18	0,14
1981	1,38	0,73	0,16	0,66	0,35	0,20	-	0,17
1982	1,71	0,49	0,58	0,65	0,69	-	-	0,08
1983	1,06	0,60	0,89	0,73	-	0,12	0,13	0,11
1984	1,92	0,75	0,78	0,87	0,77	0,12	0,22	0,21
1985	4,59	0,46	1,03	0,99	0,42	0,29	0,21	0,27
1986	1,33	0,34	0,50	0,48	-	0,12	0,10	0,10
1987	0,96	0,24	0,40	0,35	0,48	-	-	0,04
1988	0,98	0,46	0,45	0,49	0,98	0,12	-	0,13
1989	1,01	0,53	1,28	0,83	0,50	0,12	-	0,10
1990	3,20	0,88	0,72	0,96	-	0,12	0,08	0,10
1991	0,62	0,73	0,74	0,73	-	0,06	0,07	0,06
1992	1,43	0,35	0,23	0,35	-	0,06	0,08	0,06

W - White; MA - Mixed Ancestry; B - Black; T - Total;
NTDS - Neural Tube Defects; OTHER - Other Forms of spina bifida.

TABLE A.6 - THE PREVALENCE OF NTDS BY RACE IN CAPE TOWN, 1973 - 1992.
(TOTAL SPINA BIFIDA AND TOTAL NEURAL TUBE DEFECTS)

YEAR	SPINA BIFIDA				TOTAL NTD			
	W	MA	B	T	W	MA	B	T
1973	1,31	0,55	0,25	0,62	2,10	0,69	0,50	0,89
1974	0,55	0,36	0,92	0,50	2,21	0,86	1,37	1,18
1975	0,91	1,10	0,97	1,04	1,81	1,64	0,97	1,53
1976	0,62	0,80	0,24	0,65	0,93	1,04	1,19	1,05
1977	1,38	0,97	1,00	1,04	2,77	1,54	1,50	1,72
1978	1,80	0,56	0,24	0,67	2,88	0,88	0,96	1,18
1979	1,85	0,76	0,25	0,80	2,96	0,99	0,49	1,15
1980	1,46	0,72	0,36	0,72	2,55	0,94	0,54	1,04
1981	1,38	1,00	0,16	0,83	2,08	1,20	0,31	1,07
1982	2,40	0,49	0,58	0,73	3,77	0,62	0,72	1,00
1983	1,06	0,72	1,02	0,84	1,77	1,08	1,52	1,28
1984	2,68	0,87	1,00	1,07	4,22	1,56	1,00	1,63
1985	5,00	0,75	1,24	1,26	5,84	1,16	1,76	1,74
1986	1,33	0,48	0,60	0,59	1,33	0,66	0,90	0,80
1987	1,45	0,24	0,40	0,38	1,93	0,72	0,69	0,79
1988	1,95	0,58	0,45	0,63	2,93	1,22	0,82	1,18
1989	1,51	0,64	1,28	0,93	2,01	0,99	1,46	1,23
1990	3,20	1,00	0,80	1,05	3,73	1,58	0,80	1,40
1991	0,62	0,78	0,81	0,79	1,85	1,29	0,88	1,15
1992	1,43	0,40	0,30	0,41	1,43	0,58	0,61	0,63

W - White; MA - Mixed Ancestry; B - Black; T - Total;
NTDS - Neural Tube Defects

TABLE A.7 - SUMMARY TABLE OF THE TOTAL NUMBER OF NTDS AND BIRTHS BY RACE IN CAPE TOWN, 1973 - 1992.

	RACE			
	WHITE	**MIXED ANC.**	**BLACK**	**TOTAL**
Anencephaly	36	95	33	164
Encephalocele	11	21	9	41
Myelomeningocele	73	172	94	339
Other	13	37	12	62
Spina Bifida	86	209	106	401
NTDS	133	325	148	606
Births 1973-1992	51 975	308 506	155 771	516 252

NTDS - Neural Tube Defects; OTHER - Other Forms of spina bifida.
ANC. - Ancestry;

TABLE A.8 - SUMMARY TABLE OF THE OVERALL PREVALENCE OF NTDS BY RACE AND RATIO OF SPINA BIFIDA TO ANENCEPHALY IN CAPE TOWN, 1973 - 1992.

	RACE			
	WHITE	**MIXED ANC.**	**BLACK**	**ALL**
Anencephaly	0,69	0,31	0,21	0,32
Encephalocele	0,21	0,07	0,06	0,08
Myelomeningocele	1,41	0,56	0,60	0,66
Other	0,25	0,12	0,08	0,12
Spina Bifida	1,66	0,68	0,68	0,78
NTDS	2,56	1,05	0,95	1,17
Ratio Sp.Bi.:Anen	2,39	2,20	3,21	2,45

NTDS - Neural Tube Defects; OTHER - Other Forms of spina bifida;
ANC. - Ancestry; Sp.Bi. - Spina Bifida; Anen - Anencephaly.

TABLE A.9 - THE NUMBER AND PREVALENCE OF KNOWN CONCEPTIONS OF MYELOMENINGOCELE BY RACE PER MONTH AND SEASON IN CAPE TOWN, 1973 - 1992.

RACE

MONTH/ SEASON	WHITE n(PREV)	MIXED ANC. n(PREV)	BLACK n(PREV)	TOTAL n(PREV)
Jan	3(0,72)	13(0,50)	3(0,24)	20(0,47)
Feb	4(0,95)	4(0,16)	4(0,34)	13(0,32)
Mar	8(1,85)	20(0,73)	7(0,56)	35(0,79)
Apr	4(0,94)	23(0,94)	4(0,34)	32(0,79)
May	5(1,18)	13(0,55)	7(0,57)	26(0,65)
Jun	6(1,26)	12(0,52)	17(1,24)	36(0,83)
Jul	16(3,77)	24(0,96)	21(1,61)	62(1,46)
Aug	14(3,31)	10(0,38)	16(1,21)	42(0,95)
Sep	6(1,35)	10(0,39)	4(0,30)	20(0,46)
Oct	2(0,46)	17(0,75)	7(0,51)	28(0,65)
Nov	2(0,46)	8(0,30)	3(0,22)	14(0,31)
Dec	3(0,68)	18(0,69)	1(0,07)	23(0,49)
Spr	10(0,76)	35(0,45)	14(0,35)	59(0,45)
Sum	10(0,78)	35(0,45)	8(0,21)	53(0,41)
Aut	17(1,33)	56(0,74)	18(0,49)	91(0,73)
Win	36(2,72)	46(0,60)	54(1,35)	136(1,05)

n - number; PREV - prevalence; ANC. - Ancestry; Spr - Spring;
Sum - Summer; Aut - Autumn; Win - Winter.

TABLE A.10 - THE NUMBER AND PREVALENCE OF KNOWN CONCEPTIONS OF ANENCEPHALY BY RACE PER MONTH AND SEASON IN CAPE TOWN, 1973 - 1992.

	RACE			
MONTH	WHITE n(PREV)	MIXED ANC. n(PREV)	BLACK n(PREV)	TOTAL n(PREV)
Jan	1(0,24)	9(0,35)	2(0,16)	12(0,28)
Feb	2(0,48)	6(0,24)	4(0,34)	12(0,29)
Mar	2(0,46)	11(0,40)	3(0,24)	16(0,36)
Apr	1(0,24)	9(0,37)	2(0,17)	12(0,29)
May	3(0,71)	11(0,47)	3(0,24)	17(0,42)
Jun	6(1,26)	9(0,36)	6(0,44)	21(0,48)
Jul	8(1,88)	8(0,32)	2(0,15)	18(0,42)
Aug	5(1,18)	6(0,23)	2(0,15)	13(0,30)
Sep	3(1,68)	4(0,16)	4(0,30)	11(0,25)
Oct	1(0,23)	8(0,32)	1(0,07)	10(0,23)
Nov	2(0,46)	9(0,34)	2(0,15)	13(0,31)
Dec	2(0,46)	5(0,18)	2(0,14)	9(0,19)
Spr	6(0,45)	21(0,27)	7(0,17)	34(0,26)
Sum	5(0,39)	20(0,26)	8(0,21)	32(0,25)
Aut	6(0,47)	31(0,41)	8(0,22)	45(0,36)
Win	19(1,44)	23(0,30)	10(0,25)	52(0,40)

n - number; PREV - prevalence; ANC. - Ancestry; Spr - Spring;
Sum - Summer; Aut - Autumn; Win - Winter.

TABLE A.11 - THE NUMBER AND PREVALENCE OF KNOWN CONCEPTIONS OF ENCEPHALOCELES BY RACE PER MONTH AND SEASON IN CAPE TOWN, 1973 - 1992.

	RACE			
MONTH	WHITE n(PREV)	MIXED ANC. n(PREV)	BLACK n(PREV)	TOTAL n(PREV)
Jan	-(-)	2(0,08)	1(0,08)	3(0,07)
Feb	1(0,24)	1(0,04)	-(-)	2(0,05)
Mar	-(-)	2(0,07)	1(0,08)	3(0,07)
Apr	1(0,24)	3(0,12)	-(-)	4(0,10)
May	-(-)	1(0,04)	-(-)	3(0,08)
Jun	2(0,42)	4(0,16)	2(0,15)	8(0,18)
Jul	3(0,71)	3(0,12)	2(0,15)	8(0,19)
Aug	1(0,24)	1(0,04)	1(0,08)	3(0,07)
Sep	-(-)	2(0,08)	1(0,08)	3(0,07)
Oct	-(-)	2(0,08)	-(-)	2(0,05)
Nov	1(0,23)	-(-)	1(0,07)	2(0,05)
Dec	-(-)	2(0,07)	-(-)	2(0,04)
Spr	1(0,08)	4(0,05)	2(0,05)	7(0,05)
Sum	1(0,08)	5(0,06)	1(0,03)	7(0,05)
Aut	1(0,08)	6(0,08)	1(0,03)	8(0,06)
Win	6(0,45)	8(0,10)	5(0,13)	19(0,15)

n - number; PREV - prevalence; ANC. - Ancestry; Spr - Spring;
Sum - Summer; Aut - Autumn; Win - Winter.

TABLE A.12 - THE NUMBER AND PREVALENCE OF KNOWN CONCEPTIONS OF OTHER FORMS OF SPINA BIFIDA BY RACE PER MONTH AND SEASON IN CAPE TOWN, 1973 - 1992.

	RACE			
	WHITE	**MIXED ANC.**	**BLACK**	**TOTAL**
MONTH	**n(PREV)**	**n(PREV)**	**n(PREV)**	**n(PREV)**
Jan	1(0,24)	2(0,08)	1(0,08)	4(0,09)
Feb	-(-)	2(0,08)	1(0,08)	3(0,07)
Mar	1(0,23)	2(0,07)	1(0,08)	4(0,09)
Apr	1(0,24)	2(0,08)	2(0,17)	5(0,12)
May	1(0,24)	2(0,09)	-(-)	3(0,08)
Jun	3(0,63)	8(0,32)	1(0,07)	12(0,28)
Jul	3(0,71)	6(0,24)	3(0,23)	12(0,28)
Aug	2(0,47)	2(0,08)	-(-)	4(0,09)
Sep	-(-)	3(0,12)	2(0,15)	5(0,12)
Oct	1(0,23)	2(0,08)	-(-)	3(0,07)
Nov	-(-)	4(0,15)	-(-)	4(0,09)
Dec	2(0,46)	-(-)	1(0,07)	3(0,06)
Spr	1(0,08)	9(0,12)	2(0,05)	12(0,09)
Sum	3(0,24)	4(0,05)	3(0,08)	10(0,08)
Aut	3(0,23)	6(0,08)	3(0,08)	12(0,10)
Win	8(0,61)	16(0,21)	4(0,10)	28(0,22)

n - number; PREV - prevalence; ANC. - Ancestry; Spr - Spring;
Sum - Summer; Aut - Autumn; Win - Winter.

TABLE A.13 - THE NUMBER AND PREVALENCE OF KNOWN CONCEPTIONS OF SPINA BIFIDA BY RACE PER MONTH AND SEASON IN CAPE TOWN, 1973 - 1992.

	RACE			
MONTH	WHITE n(PREV)	MIXED ANC. n(PREV)	BLACK n(PREV)	TOTAL n(PREV)
Jan	4(0,95)	15(0,58)	4(0,32)	24(0,56)
Feb	4(0,95)	6(0,24)	5(0,42)	16(0,39)
Mar	9(2,08)	22(0,80)	8(0,64)	39(0,88)
Apr	5(1,18)	25(1,02)	6(0,52)	37(0,89)
May	6(1,41)	15(0,64)	7(0,57)	29(0,72)
Jun	9(1,90)	20(0,80)	18(1,32)	48(1,11)
Jul	19(4,47)	30(1,19)	24(1,84)	72(1,70)
Aug	16(3,78)	12(0,45)	16(1,21)	46(1,04)
Sep	6(1,35)	13(0,50)	6(0,46)	25(0,58)
Oct	3(0,69)	19(0,75)	7(0,51)	31(0,71)
Nov	2(0,46)	12(0,45)	3(0,22)	18(0,40)
Dec	5(1,14)	18(0,65)	2(0,14)	26(0,56)
Spr	11(0,83)	44(0,57)	16(0,40)	71(0,54)
Sum	13(1,02)	39(0,50)	11(0,28)	63(0,49)
Aut	20(1,56)	62(0,82)	21(0,58)	103(0,83)
Win	44(3,33)	62(0,81)	58(1,45)	164(1,26)

n - number; PREV - prevalence; ANC. - Ancestry; Spr - Spring;
Sum - Summer; Aut - Autumn; Win - Winter.

TABLE A.14 - THE NUMBER AND PREVALENCE OF KNOWN CONCEPTIONS OF TOTAL NTDS BY RACE PER MONTH AND SEASON IN CAPE TOWN, 1973 - 1992.

	RACE			
MONTH	**WHITE n(PREV)**	**MIXED ANC. n(PREV)**	**BLACK n(PREV)**	**TOTAL n(PREV)**
Jan	5(1,19)	26(1,00)	7(0,56)	38(0,89)
Feb	7(1,67)	13(0,53)	9(0,76)	29(0,71)
Mar	11(2,54)	35(1,27)	12(0,96)	58(1,31)
Apr	7(1,65)	37(1,51)	8(0,69)	52(1,26)
May	9(2,12)	27(1,15)	10(0,81)	46(1,15)
Jun	17(3,58)	33(1,32)	26(1,90)	76(1,75)
Jul	30(7,06)	41(1,63)	28(2,15)	99(2,33)
Aug	22(5,20)	19(0,71)	19(1,44)	60(1,36)
Sep	9(2,03)	19(0,74)	11(0,84)	39(0,90)
Oct	4(0,92)	29(1,14)	8(0,58)	41(0,94)
Nov	5(1,14)	21(0,79)	6(0,44)	32(0,72)
Dec	7(1,59)	25(0,91)	8(0,55)	40(0,86)
Spr	18(1,36)	69(0,89)	25(0,62)	112(0,85)
Sum	19(1,49)	64(0,82)	24(0,62)	107(0,82)
Aut	27(2,11)	99(1,31)	30(0,82)	156(1,25)
Win	69(5,22)	93(1,21)	73(1,83)	235(1,81)

NTDS - Neural Tube Defects; n - number; PREV - prevalence; ANC. - Ancestry; Spr - Spring; Sum - Summer; Aut - Autumn; Win - Winter.

TABLE A.15 - THE TOTAL NUMBER OF KNOWN CONCEPTIONS BY RACE PER MONTH AND SEASON IN CAPE TOWN, 1973 - 1992.

	RACE			
MONTH	**WHITE**	**MIXED ANC.**	**BLACK**	**TOTAL**
	n	**n**	**n**	**n**
Jan	4191	25930	12563	42684
Feb	4202	24772	11789	40763
Mar	4332	27519	12485	44336
Apr	4241	24526	11647	41414
May	4245	23540	12318	40103
Jun	4747	25023	13671	43441
Jul	4249	25125	13052	42426
Aug	4233	26628	13226	44087
Sep	4435	25804	13143	43382
Oct	4328	25364	13723	43415
Nov	4378	26734	13529	44641
Dec	4394	27541	14625	46560
Spr	13207	77902	40395	131504
Sum	12787	78243	38977	130007
Aut	12818	75585	36450	124853
Win	13229	76776	39949	129954

n - number; ANC. - Ancestry; Spr - Spring; Sum - Summer; Aut - Autumn; Win - Winter.

TABLE A.16 - THE NUMBER AND RATIO OF NTDS AND RACE IN CAPE TOWN, 1973 - 1992.

RACE

	WHITES		MIXED ANC.		BLACK		TOTAL	
	n		n		n		n	
	M	F(ratio)	M	F(ratio)	M	F(ratio)	M	F(ratio)
MMC	37	35(1,06)	76	94(0,81)	49	45(1,09)	162	174(0,93)
OTH.FORMS	7	6(1,17)	12	25(0,48)	6	6(1,00)	25	17(0,68)
SB.	44	41(1,03)	88	119(0,74)	55	51(1,08)	187	211(0,89)
ANEN	15	19(0,78)	38	56(0,68)	11	21(0,53)	64	96(0,67)
ENCEP	4	7(0,57)\	9	12(0,75)	4	5(0,80)	17	24(0,71)
TOTAL	64	66(0,97)	135	187(0,72)	72	76(0,95)	271	329(0,82)

n - number; ANC. - Ancestry; M - Male; F - Female;
MMC - Myelomeningocele; OTH. FORMS - Other Forms of Spina Bifida;
SB. - Spina Bifida; Anen - Anencephaly; Encep - Encephalocele.

TABLE A.17 - THE NUMBER OF MYELOMENINGOCELES BY RACE AND BIRTHS AND THE PREVALENCE PER YEAR AND 5 YEAR PERIOD OF MATERAL AGE IN CAPE TOWN, 1973 - 1992.

Age in Years	Race W	MA	B	T	Births	Prevalence
	n	n	n	n	n	
15	-	-	-	-	2014	-
16	4	7	7	18	6378	2,82
17	6	3	6	15	13455	1,11
18	5	12	16	33	22057	1,50
19	2	4	7	13	29396	0,44
20	5	10	4	19	33065	0,58
21	3	20	9	32	34764	0,92
22	4	8	6	18	35930	0,50
23	3	5	2	10	35541	0,28
24	4	10	4	18	33796	0,53
25	4	7	1	12	31810	0,38
26	4	7	2	13	30200	0,43
27	5	10	3	18	27326	0,66
28	5	11	6	22	25116	0,88
29	3	6	1	10	22669	0,44
30	2	10	3	15	22095	0,68
31	1	6	2	9	19027	0,47
32	2	4	2	8	16224	0,49
33	2	6	2	10	14252	0,70
34	2	1	2	5	12166	0,41
35	2	4	2	8	10999	0,73
36	-	4	1	5	9074	0,55
37	2	2	1	5	7324	0,68
38	2	4	2	8	6129	1,13
39	-	2	1	3	4751	0,63
40	1	1	-	2	4185	0,48
41	-	6	-	6	3032	1,98
42	-	2	2	4	2026	1,97
43	-	-	-	-	1451	-
16-20	22	36	40	98	104351	0,94
21-25	18	50	22	90	171841	0,52
26-30	19	44	15	78	127406	0,61
31-35	9	21	10	40	72668	0,55
36-40	5	13	5	23	31463	0,73
>40	-	8	2	10	6509	1,54

n - number; W - White; MA - Mixed Ancestry; B - Black

TABLE A.18 - THE NUMBER AND PREVALENCE OF NTDS PER YEAR AND 5 YEAR PERIOD OF MATERAL AGE IN CAPE TOWN, 1973 - 1992.

		TYPE OF NEURAL TUBE DEFECT					
Age in Years	**Births n**	**MMC n(Prev)**	**ANEN n(Prev)**	**ENCEPH n(Prev)**	**OTHER n(Prev)**	**SpBif n(Prev)**	**TOTAL n(Prev)**
15	2014	-(-)	1(0,50)	-(-)	-(-)	-(-)	1(0,50)
16	6378	18(2,82)	5(0,78)	-(-)	1(0,31)	20(3,14)	25(3,91)
17	13455	15(1,11)	5(0,37)	2(0,15)	2(0,15)	17(1,26)	24(1,78)
18	22057	33(1,50)	9(0,41)	1(0,05)	1(0,05)	34(1,54)	44(2,00)
19	29396	13(0,44)	8(0,27)	2(0,07)	-(-)	13(0,44)	23(0,78)
20	33065	19(0,58)	10(0,30)	1(0,03)	3(0,09)	22(0,67)	33(1,00)
21	34764	32(0,92)	18(0,52)	3(0,09)	7(0,20)	39(1,12)	60(1,73)
22	35930	18(0,50)	13(0,36)	1(0,03)	4(0,11)	22(0,61)	36(1,00)
23	35541	10(0,28)	9(0,25)	3(0,08)	5(0,14)	15(0,42)	29(0,81)
24	33796	18(0,53)	9(0,27)	3(0,09)	2(0,06)	20(0,59)	32(0,95)
25	31810	12(0,38)	9(0,28)	2(0,06)	3(0,09)	15(0,47)	26(0,82)
26	30200	13(0,43)	9(0,30)	-(-)	2(0,07)	15(0,50)	24(0,80)
27	27326	18(0,66)	4(0,15)	4(0,15)	3(0,11)	21(0,77)	29(1,07)
28	25116	22(0,88)	7(0,28)	2(0,08)	3(0,12)	25(1,00)	34(1,36)
29	22669	10(0,44)	1(0,04)	-(-)	2(0,09)	12(0,53)	13(0,57)
30	22095	15(0,68)	5(0,23)	2(0,09)	1(0,05)	16(0,72)	23(1,05)
31	19027	9(0,47)	7(0,37)	1(0,05)	2(0,11)	11(0,58)	19(1,00)
32	16224	8(0,49)	2(0,12)	2(0,12)	3(0,19)	11(0,68)	15(0,92)
33	14252	10(0,70)	4(0,28)	-(-)	4(0,28)	14(0,98)	18(1,26)
34	12166	5(0,41)	7(0,58)	2(0,16)	1(0,08)	6(0,49)	15(1,24)
35	10999	8(0,73)	1(0,09)	1(0,09)	1(0,09)	10(0,91)	12(1,09)
36	9074	5(0,55)	4(0,44)	1(0,11)	2(0,22)	7(0,77)	12(1,32)
37	7324	5(0,68)	2(0,27)	2(0,27)	2(0,27)	7(0,96)	11(1,50)
38	6129	8(1,31)	4(0,65)	-(-)	3(0,49)	11(1,80)	15(2,45)
39	4751	3(0,63)	3(0,63)	2(0,42)	-(-)	3(0,63)	8(1,68)
40	4185	2(0,48)	4(0,96)	1(0,24)	2(0,48)	4(0,960	9(2,16)
41	3032	6(1,98)	2(0,67)	-(-)	-(-)	6(1,98)	8(2,65)
42	2026	4(1,97)	1(0,49)	1(0,49)	-(-)	4(1,97)	6(2,95)
43	1451	-(-)	1(0,69)	-(-)	1(0,69)	1(0,69)	2(1,38)
16-20	104351	98(0,94)	37(0,35)	6(0,06)	8(0,08)	106(1,02)	149(1,43)
21-25	171841	90(0,52)	58(0,34)	14(0,08)	21(0,12)	111(0,65)	183(1,07)
26-30	127406	78(0,61)	26(0,20)	8(0,06)	11(0,09)	89(0,70)	123(0,97)
31-35	72668	40(0,55)	21(0,29)	6(0,08)	12(0,17)	52(0,72)	79(1,09)
36-40	31463	23(0,73)	17(0,54)	6(0,19)	9(0,29)	32(1,02)	55(1,75)
>40	6509	10(1,54)	4(0,61)	1(0,15)	1(0,15)	11(1,69)	16(2,46)

NTDS - Neural Tube Defect; n - Number; Prev - Prevalence;
MMC - Myelomeningocele; ANEN - Anencephaly; ENCEPH - Encephalocele;
OTHER - Other forms of Spina Bifida; SpBif - Spina Bidifa

TABLE A.19 - THE NUMBER AND PREVALENCE OF NTDS AND BIRTH ORDER IN CAPE TOWN, 1973 -1992

BIRTH ORDER	TOTAL BIRTHS	MMC	OTHER	SPINA BIFIDA	ANEN	TOTAL NTDS
	n	n(Prev)	n(Prev)	n(Prev)	n(Prev)	n(Prev)
1	151839	158(1,04)	14(0,09)	172(1,13)	79(0,52)	251(1,65)
2	136655	85(0,62)	24(0,18)	109(0,80)	36(0,26)	145(1,06)
3	96165	50(0,52)	9(0,09)	59(0,61)	21(0,22)	80(0,83)
4	60734	25(0,41)	4(0,07)	29(0,48)	12(0,20)	41(0,68)
5	35429	10(0,28)	5(0,14)	15(0,42)	5(0,14)	20(0,56)
6	20245	6(0,30)	-(-)	6(0,30)	3(0,15)	9(0,45)
7	10122	3(0,30)	3(0,30)	6(0,59)	5(0,49)	11(1,09)
>7	5063	2(0,40)	3(0,59)	5(0,99)	3(0,59)	8(1,58)

NTDS - Neural Tube Defects; MMC - Myelomeningocele;
OTHER - Other forms of Spina Bifida; ANEN - Anencephaly;
n - number; Prev - Prevalence.

APPENDİX 2

NAME:_____ Case ID <IDNUM>

FOLDER No: ########

SEX/RACE: #

1 - White Male
2 - White Female
3 - Coloured Male
4 - Coloured Female
5 - Asian Male
6 - Asian Female
7 - Black Male
8 - Black Female

MATERNAL AGE: ##

GRAVIDITY: ##

PARITY: ##

FAMILY HISTORY:<Y>

PREVIOUS NTD: <Y>

ANTENATAL DIAGNOSIS: #

1 - No Diagnosis/No Ultra sound(U/S)
2 - No Diagnosis/Antenatal U/S(non Peninsula Maternal and Neonatal Service - PMNS)
3 - No Diagnosis/Antenatal U/S(PMNS Obstet U/S)
4 - No Diagnosis/Antenatal U/S(PMNS Fetal U/S)
5 - Diagnosis made on U/S
6 - Diagnosis made on Other Tests

MATERNAL EDUCATION: #

1 - No Schooling
2 - Primary Schooling Only
3 - High School but not Matriculated
4 - Matriculated
5 - Tertiary Education

PATERNAL EDUCATION: #

1 - No Schooling
2 - Primary Schooling Only
3 - High School but not Matriculated
4 - Matriculated
5 - Tertiary Education

SOCIAL STATUS: #

1 - Social Classes I + II + III (Business/Professionals/Shop-Owners/ Skilled Craftsmen/White Collar Workers/Blue Collar Workers)
2 - Social Class IV (Semi-skilled)
3 - Social Class V (Unskilled)

FAMILY INCOME: ####

DATE OF BIRTH: <dd/mm/yy>

BIRTH ORIGIN: #

1 - Cape Town
2 - less than 50km from Cape Town
3 - more than 50km less than 100km from Cape Town
4 - more than 100km less than 500km from Cape Town
5 - more than 500km less than 1000km from Cape Town
6 - more than 1000km from Cape Town

BIRTH PLACE: #

1 - Home
2 - Rural Clinic
3 - Provincial Hospital
4 - PMNS Midwife Obstetric Unit (MOU)
5 - PMNS Hospital
6 - Local NonPMNS Hospital
7 - Private Hospital
8 - Other

DELIVERY MODE: #

1 - Born Before Arrival (BBA)
2 - Normal Vertex Delivery (NVD)
3 - Breech
4 - Forceps Assisted
5 - Lower Uterine Segment Caesarian Section

BIRTH MASS: ####grams

CLASSIFICATION
OF BIRTH MASS: #

1 - Underweight for Gestational Age (GA)
2 - Appropriate for GA
3 - Large for GA

BIRTH HEAD
CIRCUMFERENCE: ##.#cms

CLASSIFICATION OF HEAD
CIRCUMFERENCE(COH): #

1 - Below 10th Centile for GA
2 - Normal for GA
3 - Above 90th Centile for GA
4 - 2cms or more Above 90th Centile for GA

GROWTH
CLASSIFICATION: #

1 - Appropriate for GA
2 - Assymetrical Growth Retardation
3 - Symetrical Growth Retardation
4 - Large for GA
5 - Appropriate for GA, but COH above 90th Centile for GA
6 - Appropriate for GA, but COH below 10th Centile for GA

GESTATIONAL AGE: ##.#

MYELOMENINGOCELE
SIZE ##.#sq cms

MYELOMENINGOCELE
SITE: #

1 - Sacral
2 - Lumbo-Sacral
3 - Lumbar
4 - Thoraco-Lumbar
5 - Thoracic
6 - Cervical

CONGENITAL
ANOMALIES: #

1 - Nil, other than Myelomeningocele(MMC)
2 - Strabismus + MMC
3 - Congenital Dislocation of the Hip (CDH) + MMC
4 - Congenital Talipes Equino Varus (CTEV) + MMC
5 - CDH + CTEV + MMC
6 - Strabismus + CDH/CTEV + MMC
7 - Kyphosis/Scoliosis + MMC
8 - CDH + CTEV + Kyph/Scol + MMC
9 - Other minor/major anomaly + MMC

BIRTH SENSORI/MOTOR
LEVEL: ##

DATE OF CLOSURE: <dd/mm/yy>

DELAY IN CLOSURE: #

1 - less than 24 hours
2 - more than 24 but less than 48 hours
3 - more than 48 but less than 72 hours
4 - more than 72 but less than 168 hours
5 - more than 1/52 but less than 2/52
6 - more than 2/52 but less than 3/52
7 - more than 3/52 but less than 4/52
8 - more than 1/12

INFECTION: #

1 - No infection
2 - Superficial Wound Infection
3 - Meningitis
4 - Ventriculitis
5 - Septicaemia
6 - Pneumonia
7 - Gastroenteritis
8 - Combinations of above

DATE OF 1st VENTRICULO-PERITONEAL SHUNT: <dd/mm/yy>
DATE OF 2nd VENTRICULO-PERITONEAL SHUNT: <dd/mm/yy>
DATE OF 3rd VENTRICULO-PERITONEAL SHUNT: <dd/mm/yy>
DATE OF 4th VENTRICULO-PERITONEAL SHUNT: <dd/mm/yy>

HYDROCEPHALUS MANAGEMENT: #

1 - No Shunt Required
2 - Single VPS Required
3 - Two VPS Required
4 - Multiple VPS Required
5 - Ventricular Drain Required Only
6 - Ventricular Drain + Later VPS Required
7 - Multiple VPS + Drain
8 - Arnold Chiari Decompression
9 - Decompression + VPS +/- Drain

REASON FOR MULTIPLE SHUNTS: #

1 - Shunt Blockage
2 - Shunt Infection
3 - Shunt Blockage + Infection
4 - Other

STATUS: #

1 - Alive
2 - Died, post surgery
3 - Later death
4 - Status Unknown

DATE OF EARLY DEATH: <dd/mm/yy>

REASON FOR EARLY DEATH: #

1 - Blocked Shunt
2 - CNS Infection
3 - Other Infection
4 - Other Cause - Related to MMC
5 - Other Cause - Unrelated to MMC

DATE OF LATE DEATH: <dd/mm/yy>

REASON FOR LATE DEATH: #

1 - Blocked Shunt
2 - CNS Infection
3 - Other Infection
4 - Other Cause - Related to MMC
5 - Other Cause - Unrelated to MMC

FOLLOW UP #

1 - Completed Study
2 - Followed up in Kimberley
3 - Followed up in East London
4 - Followed up at Cecelia M. Hospital
5 - Lost To Follow Up
6 - Followed up in Port Elizabeth
7 - Followed up elsewhere

F/U ORIGIN: #

1 - Cape Town
2 - less than 50km from Cape Town
3 - more than 50km less than 100km from Cape Town
4 - more than 100km less than 500km from Cape Town
5 - more than 500km less than 1000km from Cape Town
6 - more than 1000km from Cape Town

AMBULATORY STATUS: #

1 - Not Ambulant +/- Wheelchair Bound
2 - Ambulant with BK calipers
3 - Ambulant with AK calipers
4 - Ambulant with AFO
5 - Ambulant without appliances
6 - Ambulant with other appliances
7 - Partial Ambulation

AGE AMBULANT: #

ORTHOPAEDIC SURGERY: #

1 - No surgery required
2 - Posterior Medial Release
3 - Peabody Procedure
4 - Adductor Tonotomy
5 - Combination of Procedures
6 - Other Orthopaedic Procedure

BLADDER TYPE: #

1 - Spastic
2 - Flaccid
3 - Normal

UPPER URINARY TRACT (UUT) STATUS: #

1 - Normal UUT
2 - Dilated UUT, Unilaterally
3 - Dilated UUT, Bilaterally
4 - Hydronephrosis
5 - Chronic Renal Damage

UROLOGICAL SURGERY: #

1 - No Surgery Required
2 - Required Vesicostomy
3 - Required Ureteric Reimplantation
4 - Required Urinary Diversion
5 - Combination of Procedures

BLADDER
MANAGEMENT: #

1 - Incontinent
2 - Continent on Intermittent Catheterization (IC)
3 - Continent on IC + Drugs
4 - Continent on Manual Expression
5 - Continent - Potty Trained

AGE CONTINENT: #

DEVELOPMENTAL QUOTIENTS (DQ)

DQ at 1year: ###

DQ at 2years: ###

DQ at 3years: ###

DQ at 4years: ###

GENERAL QUOTIENTS (GQ)

GQA: ###

GQB: ###

GQC: ###

GQD: ###

GQE: ###

GQF: ###

GQ5: ###

GQModified: ###

Sensori-Motor
Level at 5 years: ##

OUTCOME at 1year: ##

OUTCOME at 2years: ##

OUTCOME at 3years: ##

OUTCOME at 4years: ##

OUTCOME at 5years: ##

1 - Ambulant, Continent, N Intelligence
2 - Ambulant, Continent, Trainable
3 - Ambulant, Continent, Mentally Retarded (MR)
4 - Ambulant, Not Continent, N Intelligence
5 - Ambulant, Not Continent, Trainable
6 - Ambulant, Not Continent, MR

7 - Non Ambulant, Continent, N Intelligence
8 - Non Ambulant, Continent, Trainable
9 - Non Ambulant, Continent, MR
10 - Non Ambulant, Not Continent, N Intelligence
11 - Non Ambulant, Not Continent, Trainable
12 - Non Ambulant, Not Continent, MR

SENSORI-MOTOR
LEVEL at BIRTH: #

SENSORI-MOTOR
LEVEL at 5years: #

1 - Thoracic
2 - Lumbar 1 & 2
3 - Lumbar 3 & 4
4 - Lumbar 5
5 - Sacral 1 & 2
6 - Sacral 3, 4 & 5

SENSORI-MOTOR
LEVEL at BIRTH: ##

SENSORI-MOTOR
LEVEL at 5years: ##

1 - T 6
2 - T 7
3 - T 8
4 - T 9
5 - T10
6 - T11
7 - T12
8 - L 1
9 - L 2
10 - L 3
11 - L 4
12 - L 5
13 - S 1
14 - S 2
15 - S 3
16 - S 4
17 - S 5

–

APPENDIX 3

OUTCOME STUDY RESULTS

REC	RACE SEX	MATERNAL AGE (Yrs)	GRAVIDITY	PARITY	FAMILY HISTORY	PREVIOUS NTD	ANTENATAL DIAGNOSIS
1	4	24	1	1	N	N	1
2	3	27	4	4	N	N	1
3	3	18	1	1	N	N	1
4	8	19	3	3	N	N	3
5	3	19	1	1	N	N	5
6	7	20	3	3	N	N	1
7	3	37	5	5	N	N	1
8	1	24	2	2	N	N	1
9	3	24	3	3	N	N	1
10	7	19	1	1	N	N	1
11	7	16	1	1	N	N	5
12	4	29	3	3	N	N	1
13	3	26	3	3	N	N	1
14	1	27	3	3	N	N	3
15	3	24	3	3	N	N	1
16	2	24	1	1	N	N	1
17	4	26	2	2	N	N	1
18	1	26	3	3	N	N	2
19	4	18	1	1	N	N	1
20	4	19	2	2	N	N	1
21	3	26	2	2	N	N	1
22	1	32	1	1	N	N	2
23	3	22	2	2	N	N	1
24	1	24	1	1	N	N	2
25	4	19	1	1	N	N	1
26	4	38	8	9	N	N	3
27	8	36	4	4	N	N	1
28	4	34	6	6	N	N	1
29	4	19	1	1	N	N	1
30	3	22	1	1	N	N	1
31	2	22	1	1	N	N	1
32	3	25	2	2	N	N	1
33	3	24	2	1	N	N	1
34	1	17	1	1	N	N	1
35	3	28	4	4	N	N	1
36	1	29	4	4	N	N	1
37	3	30	5	4	N	N	1
38	3	32	6	6	N	N	1
39	4	27	3	3	N	N	1
40	4	35	5	5	N	N	1
41	4	24	3	3	N	N	1
42	8	34	6	6	N	N	1
43	2	29	3	3	N	N	2
44	7	21	1	1	N	N	1
45	7	28	3	3	N	N	1
46	8	18	1	1	N	N	1
47	3	40	3	3	N	N	1
48	8	41	6	6	N	N	1
49	2	29	1	1	N	N	1
50	8	23	2	2	N	N	1
51	1	23	1	1	N	N	1
52	2	26	2	2	N	N	1
53	7	39	1	1	N	N	1
54	8	36	5	5	N	N	1
55	7	22	2	2	N	N	1
56	8	32	3	3	N	N	1
57	4	25	1	1	N	N	1
58	3	25	2	2	N	N	1
59	3	19	1	1	N	N	1
60	4	26	3	2	N	N	5
61	4	21	1	1	N	N	1
62	3	42	3	3	N	N	1
63	2	26	4	4	N	N	1
64	3	28	4	4	N	N	1
65	7	26	3	3	N	N	1
66	7	19	1	1	N	N	1
67	4	25	1	1	Y	N	2
68	4	26	2	2	N	N	3

REC	RACE SEX	MATERNAL AGE (Yrs)	GRAVIDITY	PARITY	FAMILY HISTORY	PREVIOUS NTD	ANTENATAL DIAGNOSIS
69	1	26	2	2	N	N	1
70	3	24	2	2	N	N	1
71	2	25	1	1	N	N	3
72	3	20	1	1	N	N	1
73	4	26	2	2	N	N	1
74	2	25	1	1	N	N	2
75	1	22	2	2	N	N	2
76	3	28	3	3	N	N	1
77	3	31	3	3	N	N	2
78	8	36	5	5	N	N	1
79	8	26	3	3	N	N	1
80	7	23	1	1	N	N	1
81	7	17	1	1	N	N	1
82	7	27	5	5	Y	Y	1
83	8	33	4	4	N	N	1
84	3	20	1	1	N	N	3
85	4	20	1	1	N	N	1
86	3	26	2	2	N	N	1
87	3	25	3	3	N	N	1
88	8	37	7	7	N	N	1
89	3	17	1	1	N	N	1
90	2	24	2	2	N	N	1
91	4	30	2	2	N	N	3
92	2	23	1	1	N	N	1
93	4	32	4	4	N	N	1
94	3	42	6	6	N	N	5
95	1	32	3	3	N	N	1
96	4	21	1	1	N	N	1
97	1	24	3	3	N	N	1
98	3	35	1	1	N	N	2
99	3	34	4	4	N	N	1
100	3	24	1	1	N	N	1
101	4	22	1	1	N	N	3
102	8	23	2	2	N	N	1
103	8	37	5	5	N	N	1
104	8	25	2	2	N	N	1
105	7	32	1	1	N	N	1
106	3	55	10	10	N	N	1
107	1	27	1	1	N	N	1
108	3	36	2	2	N	N	1
109	1	26	1	1	N	N	1
110	3	32	5	3	N	N	2
111	1	34	3	3	N	N	1
112	4	30	3	3	N	N	2
113	4	35	1	1	N	N	1
114	4	24	2	2	N	N	1
115	4	28	3	3	N	N	1
116	4	19	1	1	N	N	1
117	3	34	3	2	N	N	1
118	8	24	1	1	N	N	1

OUTCOME STUDY RESULTS

REC	MATERNAL EDUCATION	PATERNAL EDUCATION	SOCIAL STATUS	FAMILY INCOME(R)	DATE OF BIRTH	BIRTH ORIGIN	PLACE OF BIRTH
1	1	1	3	100	12/02/79	4	3
2	1	1	3	50	27/10/81	1	1
3	1	1	3	60	19/05/81	5	1
4	2	2	3	120	30/04/83	1	5
5	3	3	2	120	22/02/84	1	5
6	1	1	3	90	12/11/84	1	4
7	1	1	3	32	30/01/81	4	3
8	4	4	1	2500	02/03/82	1	6
9	3	3	1	300	08/08/83	1	5
10	2	1	3	60	15/05/82	1	5
11	3	2	3	60	09/07/84	1	5
12	5	4	1	476	26/07/84	3	2
13	1	1	3	350	01/02/79	1	5
14	4	3	1	1790	26/05/79	1	5
15	1	1	3	20	03/06/79	3	1
16	4	4	1	2500	05/12/79	1	7
17	3	3	1	500	02/08/79	5	8
18	3	3	1	1000	12/12/79	1	6
19	1	1	3	40	19/02/80	4	3
20	2	1	3	132	26/04/80	1	5
21	3	3	2	200	30/04/81	2	5
22	4	5	1	3000	22/08/80	1	5
23	2	1	3	440	20/10/80	4	3
24	4	4	1	4000	07/04/80	1	5
25	3	3	3	433	19/04/80	1	5
26	2	2	3	300	30/12/80	1	5
27	2	2	2	450	28/09/80	5	3
28	1	1	3	75	28/10/80	4	1
29	3	3	2	450	08/07/81	3	3
30	3	3	2	390	06/11/81	1	4
31	4	4	1	1700	05/05/81	4	7
32	3	4	1	2800	21/12/81	1	5
33	3	2	3	400	29/05/81	4	3
34	4	4	1	2500	12/05/81	1	5
35	3	2	2	254	27/06/81	1	4
36	4	4	1	1750	04/09/81	4	3
37	3	3	1	350	04/03/81	1	1
38	1	1	3	60	21/04/81	5	1
39	2	3	3	471	09/01/81	1	4
40	1	1	3	120	19/04/83	4	1
41	2	3	2	680	29/09/81	1	5
42	3	3	2	250	17/08/81	5	3
43	4	4	1	3000	08/12/81	5	3
44	1	1	3	108	08/10/81	1	5
45	1	2	3	280	23/11/81	5	2
46	2	2	3	50	05/10/81	5	3
47	2	2	2	320	03/08/81	4	3
48	1	1	3	120	07/12/81	5	1
49	5	4	1	5000	29/08/82	1	5
50	2	2	2	180	12/05/82	5	2
51	3	3	1	1000	08/03/82	3	3
52	4	4	1	2500	26/08/82	5	3
53	3	3	1	900	15/11/82	6	3
54	1	1	3	49	12/07/82	5	1
55	2	2	3	50	11/09/82	5	3
56	2	2	3	200	30/09/82	5	3
57	3	3	1	1600	03/01/82	1	5
58	1	1	3	40	07/10/82	4	3
59	2	2	3	147	15/01/82	2	3
60	2	3	2	350	27/12/82	3	2
61	2	3	1	368	14/10/82	1	5
62	1	1	3	100	31/08/82	4	1
63	3	3	2	1800	12/05/82	1	5
64	1	1	3	120	04/09/82	4	3
65	2	3	3	90	23/03/82	5	3
66	1	1	3	200	14/04/82	5	3
67	4	3	1	680	07/12/83	1	5
68	4	4	1	1600	04/09/83	1	5

REC	MATERNAL EDUCATION	PATERNAL EDUCATION	SOCIAL STATUS	FAMILY INCOME(R)	DATE OF BIRTH	BIRTH ORIGIN	PLACE OF BIRTH
69	3	4	1	2000	02/02/83	5	3
70	2	2	3	350	01/11/83	5	3
71	4	4	1	3000	05/02/83	1	5
72	4	4	1	450	25/10/83	3	3
73	3	4	1	1000	10/04/83	1	5
74	4	5	1	3500	18/04/83	1	5
75	4	4	1	2670	01/09/83	5	3
76	3	3	1	520	05/02/83	1	4
77	1	1	3	130	15/07/80	1	6
78	2	2	3	147	04/05/83	5	1
79	3	2	3	175	04/02/83	5	8
80	1	1	3	60	12/03/83	5	3
81	3	3	3	390	30/05/83	1	4
82	2	2	3	200	27/02/83	6	3
83	1	1	3	75	18/08/83	5	1
84	4	3	1	600	10/01/83	1	4
85	2	2	3	100	12/07/83	4	3
86	3	4	1	678	13/11/83	5	3
87	2	2	3	100	01/04/83	4	1
88	1	1	3	60	19/09/83	5	2
89	2	3	3	180	06/02/84	4	3
90	4	4	1	1900	05/12/84	1	5
91	3	3	1	1800	03/04/84	1	5
92	4	3	1	3000	31/07/84	4	3
93	1	1	3	150	17/03/84	5	1
94	2	3	2	600	15/08/84	1	5
95	3	3	2	900	28/09/84	4	3
96	2	2	3	43	28/11/84	5	3
97	4	3	1	900	20/03/84	5	3
98	4	3	1	2316	12/04/84	1	7
99	2	1	3	400	03/12/84	1	4
100	1	1	2	60	23/10/84	4	3
101	3	3	2	1700	01/10/84	1	4
102	1	1	3	275	11/10/84	5	1
103	2	3	3	75	05/04/84	5	3
104	2	2	3	250	07/04/84	5	3
105	5	4	1	2000	01/02/84	1	6
106	1	1	3	250	02/03/84	4	1
107	5	5	1	3000	22/05/84	1	7
108	1	1	3	195	20/02/84	1	4
109	3	4	1	1500	21/06/82	5	3
110	4	4	1	4000	14/01/81	1	6
111	4	4	1	2680	03/01/81	1	6
112	2	2	3	750	11/05/81	1	6
113	4	4	1	1500	27/06/80	4	3
114	1	1	3	65	09/06/79	4	1
115	3	3	2	350	08/04/79	5	3
116	2	2	3	175	29/08/79	1	5
117	2	2	3	40	21/04/79	4	3
118	1	1	3	300	26/08/84	5	3

OUTCOME STUDY RESULTS

REC	MODE of DELIVERY	BIRTH MASS grams	CLASSIFICATION BIRTH MASS	BIRTH COH cms	CLASSIFICATION COH	GROWTH CLASSIF.	GESTATIONAL AGE weeks
1	2	2800	2	34.5	2	1	40.0
2	1	1240	1	26.0	1	3	32.0
3	1	2600	2	34.0	3	5	36.0
4	2	2890	2	34.5	2	1	40.0
5	2	4000	3	36.0	2	4	40.0
6	2	2540	2	32.0	2	1	38.0
7	2	2360	2	32.0	2	1	37.0
8	2	2900	2	32.5	1	6	40.0
9	5	3360	2	34.5	2	1	38.0
10	2	4300	3	39.5	4	4	40.0
11	5	2720	2	38.0	4	5	38.0
12	2	3890	2	34.5	2	1	40.0
13	4	3560	2	35.5	2	1	40.0
14	2	4120	3	35.8	2	4	40.0
15	1	2340	2	33.0	2	1	36.0
16	5	4580	3	34.0	2	4	40.0
17	2	3700	2	35.0	2	1	40.0
18	5	2980	2	38.0	3	5	40.0
19	2	3420	2	34.0	2	1	40.0
20	5	3320	2	36.5	3	5	28.0
21	4	2473	2	32.0	2	1	38.0
22	4	3250	2	30.8	1	6	37.0
23	2	3360	2	36.0	2	1	40.0
24	2	3340	2	34.0	2	1	40.0
25	5	4100	3	37.5	4	4	36.0
26	2	3100	2	35.5	2	1	38.0
27	2	2620	2	34.5	2	1	38.0
28	1	2650	1	33.1	2	2	40.0
29	2	3200	2	34.0	2	1	40.0
30	2	2880	2	36.0	2	1	40.0
31	2	3400	2	34.0	2	1	38.6
32	2	3260	2	35.0	2	1	39.0
33	2	3000	2	35.0	2	1	39.0
34	2	3100	2	40.1	4	5	40.0
35	2	2500	2	33.0	2	1	38.0
36	2	3660	2	34.5	2	1	40.0
37	1	1620	1	29.0	1	3	35.0
38	1	2100	2	31.0	2	1	36.0
39	2	2420	1	33.0	2	2	39.0
40	1	2400	2	30.5	1	6	37.0
41	2	3100	2	33.0	2	1	40.0
42	2	2950	2	35.0	2	1	39.0
43	2	3680	2	35.0	2	1	40.0
44	2	2720	2	34.0	2	1	38.5
45	2	3080	2	36.2	3	5	38.0
46	2	2250	1	33.5	2	2	40.0
47	2	4620	3	35.0	2	4	40.0
48	1	2000	2	32.5	2	1	36.0
49	2	3140	2	34.5	2	1	38.6
50	2	2400	1	30.0	1	3	40.0
51	2	4220	3	36.0	2	4	40.0
52	2	2720	2	33.5	2	1	38.0
53	5	3150	2	35.5	2	1	38.0
54	1	2450	1	34.4	2	2	40.0
55	2	3000	2	35.0	2	1	40.0
56	2	3900	2	38.0	3	5	40.0
57	5	3400	2	37.0	4	5	37.0
58	2	3600	2	35.5	2	1	38.0
59	2	2540	2	30.5	2	1	36.0
60	2	2680	2	34.0	2	1	38.0
61	5	2850	2	36.0	3	5	38.2
62	1	1960	1	29.8	1	3	36.8
63	2	3000	2	33.0	2	1	37.8
64	2	2580	1	34.0	2	2	40.0
65	2	3200	2	34.5	2	1	40.0
66	2	3540	2	36.0	3	5	39.0
67	2	3180	2	35.0	2	1	38.2
68	2	3180	2	32.0	1	5	40.0

REC	MODE of DELIVERY	BIRTH MASS grams	CLASSIFICATION BIRTH MASS	BIRTH COH cms	CLASSIFICATION COH	GROWTH CLASSIF.	GESTATIONAL AGE weeks
69	2	4120	3	35.0	2	4	40.0
70	2	3820	2	33.5	2	1	40.0
71	2	2900	2	33.0	2	1	38.5
72	2	2500	2	32.0	2	1	37.6
73	4	4000	3	36.0	2	4	40.0
74	2	2870	2	33.0	2	1	38.6
75	5	4700	3	36.0	2	4	40.0
76	2	2975	2	33.0	2	1	39.5
77	4	2680	1	34.5	2	1	40.0
78	1	2600	1	36.0	2	2	40.0
79	2	3300	2	37.0	3	5	40.0
80	2	3200	2	34.5	2	1	40.0
81	3	2900	2	35.5	2	1	38.5
82	2	3890	2	35.8	2	1	39.6
83	1	2120	2	32.5	2	1	36.0
84	2	2460	2	31.5	2	1	38.0
85	2	2870	2	36.0	2	1	40.0
86	2	3320	2	35.5	2	1	40.0
87	1	3590	2	37.0	3	5	39.0
88	2	3150	2	37.0	3	5	40.0
89	2	3300	2	36.0	2	1	38.0
90	5	3100	2	34.0	2	1	39.0
91	5	3500	2	35.0	2	1	38.6
92	5	3250	2	34.5	2	1	40.0
93	1	2280	2	34.5	2	1	36.0
94	5	2520	2	36.0	3	5	37.0
95	2	2100	2	33.5	2	1	36.0
96	2	2750	2	38.0	3	5	40.0
97	2	3800	3	36.5	3	4	39.0
98	4	1500	1	29.0	1	3	36.0
99	3	2100	2	31.0	2	1	36.0
100	2	1800	1	29.0	1	3	36.0
101	2	3600	2	32.0	1	6	40.0
102	1	2450	1	34.5	2	2	40.0
103	2	2600	2	34.0	2	1	38.5
104	5	4000	3	38.0	3	4	39.5
105	2	3800	2	36.5	2	1	40.0
106	1	2500	2	42.5	4	5	38.5
107	5	2940	2	34.0	2	1	38.6
108	2	3400	2	35.0	2	1	39.0
109	5	4240	3	36.5	2	4	40.0
110	2	3984	2	36.5	2	1	40.0
111	5	3540	2	36.5	3	5	38.0
112	2	2680	2	35.0	2	1	38.0
113	2	2210	1	32.5	2	2	38.0
114	1	2960	2	36.0	2	1	40.0
115	2	3480	2	36.5	2	1	40.0
116	5	2500	2	31.5	2	1	38.0
117	2	3730	2	38.0	3	5	40.0
118	2	2650	2	34.0	2	1	37.0

OUTCOME STUDY RESULTS

REC	SIZE OF MMC sq. cm.	SITE OF MMC	CONGENITAL ANOMALIES	SENSORI MOTOR LEVEL @ BIRTH	DATE OF CLOSURE	CLOSURE DELAY	INFECTION
1	25.0	3	1	L 5	20/02/79	5	2
2	12.0	2	1	S 4	28/10/81	1	1
3	6.0	1	1	L 5	11/06/81	7	4
4	20.0	4	1	L 2	01/05/83	1	1
5	20.0	3	1	L 4	23/02/84	1	4
6	16.0	2	1	L 4	13/11/84	2	2
7	24.0	2	2	L 5	04/02/81	4	2
8	12.0	2	2	L 3	08/03/82	4	2
9	20.0	2	1	L 2	09/08/83	1	1
10	12.0	2	2	L 5	16/05/82	1	3
11	16.0	3	1	T12	10/07/84	1	1
12	9.0	4	7	L 5	26/07/84	1	1
13	20.0	3	4	L 5	03/02/79	2	1
14	24.0	3	2	L 5	26/05/79	1	1
15	8.0	2	4	S 2	08/06/79	4	2
16	6.0	2	2	S 1	05/12/79	1	1
17	10.0	1	1	S 4	22/10/79	8	1
18	20.0	4	4	L 2	28/02/80	8	1
19	8.0	2	1	S 4	21/02/80	3	1
20	28.0	3	1	S 1	26/04/80	1	1
21	8.0	3	7	L 3	21/05/81	6	4
22	4.0	2	7	S 1	22/08/80	1	3
23	10.0	2	1	S 1	21/10/80	1	1
24	8.0	2	1	S 2	07/04/80	1	1
25	15.0	1	4	S 4	19/04/80	1	1
26	12.0	4	2	L 3	30/12/80	1	1
27	6.0	3	1	L 4	23/10/80	7	4
28	16.0	3	4	L 5	04/11/80	4	1
29	8.0	2	2	L 5	09/07/81	2	1
30	12.0	2	1	S 2	09/11/81	2	1
31	20.0	2	1	S 2	05/05/81	1	1
32	9.0	3	1	S 2	21/12/81	1	1
33	6.0	1	3	S 1	30/07/81	8	2
34	20.0	2	1	S 3	12/05/81	1	1
35	8.0	2	1	L 5	28/06/81	1	1
36	25.0	3	2	S 2	08/09/81	4	1
37	2.0	3	1	S 4	09/03/81	4	2
38	8.0	3	4	L 2	24/04/81	4	2
39	14.0	1	1	S 4	10/01/81	1	1
40	4.0	3	2	L 5	21/04/83	2	1
41	8.0	1	1	S 3	01/10/81	2	1
42	80.0	3	1	L 3	24/08/81	5	2
43	16.0	2	4	L 3	08/12/81	1	8
44	16.0	2	9	S 1	08/10/81	1	1
45	16.0	3	1	L 4	26/11/81	4	3
46	11.0	2	9	L 4	12/10/81	5	1
47	8.0	2	1	S 5	06/08/81	4	1
48	9.0	4	1	L 2	14/12/81	5	1
49	20.0	5	4	T12	02/12/82	8	1
50	2.0	5	1	T 7	27/05/82	6	2
51	10.0	3	2	S 1	11/03/82	3	2
52	12.0	2	1	S 3	27/08/82	2	1
53	12.0	3	1	L 2	25/04/83	8	4
54	24.0	3	1	L 3	13/09/82	8	2
55	16.0	3	1	S 3	30/09/82	6	1
56	30.0	2	1	L 5	01/11/82	8	1
57	18.0	2	1	S 3	04/01/82	1	1
58	24.0	3	1	L 3	07/10/82	1	1
59	6.0	2	4	S 4	21/01/82	4	1
60	12.0	3	1	L 5	28/12/82	2	1
61	25.0	3	5	L 2	14/10/82	1	1
62	48.0	3	4	L 5	01/09/82	2	1
63	6.0	2	1	S 1	13/05/82	1	1
64	12.0	5	4	T11	19/05/83	8	2
65	4.0	2	1	L 4	29/03/82	4	3
66	24.0	2	1	L 5	22/04/82	5	2
67	20.0	2	1	S 3	07/12/83	1	1
68	6.0	2	6	S 2	05/09/83	2	1

REC	SIZE OF MMC sq. cm.	SITE OF MMC	CONGENITAL ANOMALIES	SENSORI MOTOR LEVEL @ BIRTH	DATE OF CLOSURE	CLOSURE DELAY	INFECTION
69	10.0	3	2	L 5	04/02/83	3	1
70	49.0	2	1	L 5	10/11/83	5	1
71	9.0	2	1	S 4	05/02/83	1	1
72	5.0	2	1	S 3	25/10/83	1	1
73	4.0	2	1	S 4	30/05/83	8	1
74	6.0	3	1	L 5	18/04/83	1	1
75	4.0	2	1	S 5	02/09/83	2	1
76	12.0	2	1	S 2	05/02/83	1	1
77	16.0	2	4	S 1	26/07/80	5	1
78	30.0	2	1	L 2	16/05/83	5	2
79	16.0	3	3	L 3	07/03/83	8	1
80	12.0	3	9	L 1	14/04/83	8	2
81	20.0	3	4	L 2	30/05/83	1	2
82	16.0	6	1	T 6	01/03/83	2	1
83	6.0	3	4	L 3	07/09/83	6	2
84	15.0	4	1	T12	11/01/83	2	1
85	12.0	2	1	S 2	14/07/83	2	1
86	18.0	2	1	S 2	17/11/83	4	1
87	9.0	2	1	L 4	14/04/83	5	1
88	9.0	5	1	T 8	01/12/83	8	1
89	4.0	2	9	L 5	27/02/84	5	1
90	9.0	4	8	L 3	07/12/84	2	1
91	6.0	3	3	L 3	03/04/84	1	1
92	9.0	3	1	L 3	03/08/84	3	2
93	9.0	2	1	L 3	21/03/84	4	4
94	12.0	3	5	L 3	15/08/84	1	1
95	6.0	1	1	S 2	18/10/84	6	1
96	12.0	4	1	L 1	06/12/84	5	2
97	9.0	1	1	S 3	29/03/84	5	4
98	9.0	2	9	S 3	12/04/84	1	3
99	6.0	1	1	S 4	03/12/84	1	1
100	9.0	3	1	L 5	25/10/84	3	2
101	6.0	1	9	S 4	02/10/84	2	1
102	12.0	4	1	L 4	12/11/84	8	1
103	12.0	1	1	S 2	06/04/84	2	1
104	25.0	3	4	L 5	16/04/84	5	1
105	8.0	2	4	S 1	01/02/84	1	1
106	20.0	4	1	L 2	02/04/84	7	4
107	6.0	1	1	S 5	23/05/84	1	1
108	8.0	2	1	S 1	20/02/84	1	1
109	12.0	4	5	L 3	30/06/82	5	2
110	20.0	3	1	L 4	15/01/81	1	1
111	8.0	4	1	L 1	05/01/81	2	1
112	10.0	2	4	L 5	23/06/81	8	2
113	20.0	2	1	L 5	15/07/80	6	2
114	25.0	2	4	L 1	30/06/79	7	4
115	12.0	3	1	L 5	18/04/79	5	2
116	9.0	2	3	L 5	30/08/79	2	3
117	10.0	2	1	L 2	27/08/79	8	4
118	9.0	4	9	L 4	05/09/84	5	2

OUTCOME STUDY RESULTS

REC	DATE 1st SHUNT	DATE 2nd SHUNT	DATE 3rd SHUNT	DATE 4th SHUNT	HYDROCEPHALUS MANAGEMENT	REASON FOR REPEAT SHUNTS
1	.	.	.	.	8	.
2	.	.	.	.	1	.
3	.	.	.	.	5	.
4	.	.	.	.	1	.
5	.	.	.	.	5	.
6	26/11/84	.	.	.	2	.
7	13/08/81	.	.	.	2	.
8	01/04/82	03/05/82	.	.	3	1
9	22/08/83	31/10/83	19/01/84	.	4	3
10	07/06/82	22/07/82	.	.	7	3
11	23/07/84	.	.	.	2	.
12	14/08/84	28/08/84	13/02/86	.	4	1
13	05/02/79	.	.	.	2	.
14	11/06/79	26/06/79	.	.	3	1
15	14/06/79	07/05/84	.	.	3	1
16	20/12/79	.	.	.	2	.
17	.	.	.	.	1	.
18	18/02/80	01/04/80	01/09/80	02/10/80	3	2
19	.	.	.	.	1	.
20	15/05/80	.	.	.	2	.
21	23/06/81	02/04/83	.	.	3	1
22	28/08/80	09/02/81	.	.	3	2
23	.	.	.	.	1	.
24	17/04/80	.	.	.	2	.
25	08/05/80	.	.	.	2	.
26	19/02/81	28/09/81	.	.	3	2
27	27/11/80	.	.	.	2	.
28	20/11/80	.	.	.	2	.
29	16/07/81	.	.	.	2	.
30	23/11/81	.	.	.	2	.
31	25/05/81	.	.	.	2	.
32	.	.	.	.	1	.
33	09/11/81	17/03/82	.	.	3	2
34	04/06/81	17/09/81	.	.	3	1
35	13/07/81	.	.	.	2	.
36	04/10/81	.	.	.	2	.
37	.	.	.	.	1	.
38	09/06/81	05/04/82	.	.	3	1
39	.	.	.	.	1	.
40	02/05/83	16/05/83	15/09/83	.	4	1
41	.	.	.	.	1	.
42	05/10/81	.	.	.	2	.
43	.	.	.	.	5	.
44	02/11/81	.	.	.	2	.
45	07/12/81	.	.	.	2	.
46	07/12/81	.	.	.	2	.
47	.	.	.	.	1	.
48	31/12/81	.	.	.	2	.
49	30/09/82	06/12/82	07/12/82	.	4	3
50	07/06/82	.	.	.	2	.
51	15/04/82	.	.	.	2	.
52	.	.	.	.	1	.
53	31/03/83	03/04/83	10/08/83	.	4	2
54	.	.	.	.	1	.
55	.	.	.	.	1	.
56	11/11/82	.	.	.	2	.
57	04/03/82	15/03/82	.	.	3	1
58	18/10/82	.	.	.	2	.
59	.	.	.	.	1	.
60	10/01/83	.	.	.	2	.
61	28/10/82	13/12/82	.	.	3	1
62	14/09/82	.	.	.	1	.
63	07/06/82	16/08/82	21/01/83	27/01/83	7	3
64	04/10/82	28/10/82	.	.	3	1
65	05/04/82	.	.	.	2	.
66	.	.	.	.	1	.
67	15/12/83	02/02/84	.	.	7	2
68	18/12/83	.	.	.	2	.

REC	DATE 1st SHUNT	DATE 2nd SHUNT	DATE 3rd SHUNT	DATE 4th SHUNT	HYDROCEPHALUS MANAGEMENT	REASON FOR REPEAT SHUNTS
69	07/03/83	.	.	.	2	.
70	.	.	.	.	1	.
71	07/02/83	03/03/83	.	.	3	3
72	.	.	.	.	1	.
73	.	.	.	.	1	.
74	05/05/83	04/08/83	22/09/83	.	4	1
75	22/09/83	.	.	.	2	.
76	14/02/83	.	.	.	2	.
77	.	.	.	.	1	.
78	.	.	.	.	1	.
79	28/03/83	19/11/83	30/11/83	06/12/83	6	2
80	04/07/83	10/11/83	19/11/83	06/12/84	3	3
81	25/08/83	08/09/83	31/10/83	11/11/83	4	3
82	.	.	.	.	1	.
83	22/09/83	.	.	.	2	.
84	27/01/83	.	.	.	2	.
85	21/07/83	.	.	.	2	.
86	01/12/83	.	.	.	2	.
87	11/08/83	.	.	.	2	.
88	12/12/83	.	.	.	2	.
89	05/03/84	15/03/84	.	.	3	1
90	19/12/84	09/08/85	.	.	3	1
91	16/04/84	.	.	.	2	.
92	07/08/84	01/09/84	28/02/87	12/08/88	4	3
93	19/04/84	04/06/84	.	.	3	1
94	27/12/84	.	.	.	2	.
95	.	.	.	.	1	.
96	13/12/84	.	.	.	2	.
97	26/04/84	.	.	.	6	.
98	30/04/84	26/05/84	05/01/86	.	7	2
99	.	.	.	.	1	.
100	08/11/84	.	.	.	2	.
101	18/02/84	.	.	.	2	.
102	01/11/84	.	.	.	2	.
103	19/04/84	.	.	.	2	.
104	29/11/84	.	.	.	2	.
105	20/02/84	09/04/84	01/09/85	03/10/85	7	3
106	12/04/84	.	.	.	2	.
107	08/11/84	.	.	.	2	.
108	.	.	.	.	1	.
109	.	.	.	.	1	.
110	31/03/81	22/04/82	30/08/84	.	4	1
111	10/09/81	29/08/84	06/09/84	.	3	1
112	22/07/81	.	.	.	2	.
113	28/06/80	.	.	.	2	.
114	20/07/79	28/08/79	.	.	3	1
115	30/04/79	.	.	.	2	.
116	21/08/79	.	.	.	2	.
117	13/08/79	23/04/81	04/05/81	14/05/81	7	3
118	.	.	.	.	1	.

OUTCOME STUDY RESULTS

REC	STATUS	DATE OF EARLY DEATH	EARLY DEATH	DATE OF LATE DEATH	LATER DEATH	FOLLOWUP	ORIGIN FOR FOLLOWUP
1	2	21/02/79	4	.	.	.	.
2	2	29/10/81	4	.	.	.	.
3	2	17/06/81	2	.	.	.	.
4	2	03/05/83	3	.	.	.	.
5	2	25/02/84	4	.	.	.	.
6	2	.	.	23/12/84	2	.	.
7	3	.	.	12/02/82	1	.	.
8	3	.	.	08/06/82	3	.	.
9	3	.	.	31/01/84	3	.	.
10	3	.	.	08/09/82	2	.	.
11	3	.	.	29/11/85	3	.	.
12	3	.	.	07/07/86	4	.	.
13	1	.	.	.	.	1	1
14	1	.	.	.	.	1	1
15	1	.	.	.	.	1	3
16	1	.	.	.	.	1	1
17	1	.	.	.	.	1	5
18	1	.	.	.	.	1	1
19	1	.	.	.	.	1	4
20	1	.	.	.	.	1	1
21	1	.	.	.	.	1	2
22	1	.	.	.	.	1	1
23	1	.	.	.	.	1	4
24	1	.	.	.	.	1	1
25	1	.	.	.	.	1	1
26	1	.	.	.	.	1	1
27	1	.	.	.	.	2	5
28	1	.	.	.	.	5	4
29	1	.	.	.	.	1	3
30	1	.	.	.	.	1	1
31	1	.	.	.	.	1	4
32	1	.	.	.	.	1	6
33	1	.	.	.	.	1	4
34	1	.	.	.	.	1	1
35	1	.	.	.	.	1	1
36	1	.	.	.	.	1	4
37	1	.	.	.	.	1	1
38	1	.	.	.	.	1	5
39	1	.	.	.	.	1	1
40	1	.	.	.	.	1	4
41	1	.	.	.	.	1	1
42	1	.	.	.	.	5	5
43	1	.	.	.	.	1	5
44	1	.	.	.	.	1	1
45	1	.	.	.	.	1	5
46	1	.	.	.	.	1	5
47	1	.	.	.	.	5	4
48	1	.	.	.	.	5	5
49	1	.	.	.	.	1	2
50	1	.	.	.	.	5	5
51	1	.	.	.	.	1	3
52	1	.	.	.	.	1	5
53	1	.	.	.	.	1	6
54	1	.	.	.	.	5	5
55	1	.	.	.	.	2	5
56	1	.	.	.	.	1	5
57	4	.	.	.	.	5	1
58	1	.	.	.	.	5	4
59	1	.	.	.	.	1	2
60	1	.	.	.	.	1	3
61	1	.	.	.	.	1	1
62	1	.	.	.	.	1	4
63	1	.	.	.	.	1	1
64	1	.	.	.	.	1	4
65	1	.	.	.	.	2	5
66	1	.	.	.	.	5	5
67	1	.	.	.	.	1	1
68	1	.	.	.	.	1	1

REC	STATUS	DATE OF EARLY DEATH	EARLY DEATH	DATE OF LATE DEATH	LATER DEATH	FOLLOWUP	ORIGIN FOR FOLLOWUP
69	1	.	.	.	.	3	5
70	1	.	.	.	.	2	5
71	1	.	.	.	.	1	1
72	1	.	.	.	.	1	3
73	1	.	.	.	.	1	2
74	1	.	.	.	.	1	1
75	1	.	.	.	.	3	5
76	1	.	.	.	.	1	1
77	1	.	.	.	.	1	1
78	1	.	.	.	.	2	5
79	1	.	.	.	.	5	5
80	1	.	.	.	.	4	5
81	1	.	.	.	.	1	1
82	1	.	.	.	.	1	6
83	1	.	.	.	.	5	5
84	1	.	.	.	.	1	1
85	1	.	.	.	.	1	1
86	1	.	.	.	.	1	5
87	1	.	.	.	.	1	4
88	1	.	.	.	.	2	5
89	1	.	.	.	.	1	4
90	1	.	.	.	.	1	1
91	1	.	.	.	.	1	4
92	1	.	.	.	.	1	4
93	1	.	.	.	.	1	5
94	1	.	.	.	.	1	1
95	1	.	.	.	.	1	4
96	1	.	.	.	.	2	5
97	1	.	.	.	.	1	5
98	1	.	.	.	.	1	1
99	1	.	.	.	.	1	1
100	1	.	.	.	.	5	4
101	1	.	.	.	.	1	1
102	1	.	.	.	.	3	5
103	1	.	.	.	.	3	5
104	1	.	.	.	.	5	5
105	1	.	.	.	.	1	1
106	1	.	.	.	.	1	4
107	1	.	.	.	.	1	1
108	1	.	.	.	.	1	1
109	1	.	.	.	.	1	1
110	1	.	.	.	.	1	1
111	1	.	.	.	.	1	1
112	1	.	.	.	.	1	1
113	1	.	.	.	.	1	4
114	1	.	.	.	.	1	4
115	1	.	.	.	.	1	5
116	1	.	.	.	.	1	1
117	1	.	.	.	.	5	4
118	1	.	.	.	.	1	5

OUTCOME STUDY RESULTS

REC	AMBULATORY STATUS	AGE AMBULANT	ORTHOPAEDIC SURGERY	BLADDER TYPE	UPPER URINARY TRACT STATUS	UROLOGICAL SURGERY	BLADDER MANAGE-MENT	AGE CONTI-NENT
1	.	.	.	.	.	.	.	.
2	.	.	.	.	.	.	.	.
3	.	.	.	.	.	.	.	.
4	.	.	.	.	.	.	.	.
5	.	.	.	.	.	.	.	.
6	.	.	.	.	.	.	.	.
7	.	.	.	.	.	.	.	.
8	.	.	.	.	.	.	.	.
9	.	.	.	.	.	.	.	.
10	.	.	.	.	.	.	.	.
11	.	.	.	.	.	.	.	.
12	.	.	.	.	.	.	.	.
13	2	2	2	2	1	1	1	.
14	2	3	6	1	2	3	1	.
15	5	3	3	1	1	1	1	.
16	5	3	1	1	2	5	1	.
17	5	1	2	1	2	1	3	3
18	1	.	5	1	1	1	1	.
19	5	1	1	1	2	3	1	.
20	2	4	1	2	1	1	3	3
21	1	.	1	1	2	1	1	.
22	2	3	2	1	1	1	3	4
23	5	3	1	1	1	1	1	.
24	5	3	2	3	1	1	3	4
25	5	2	1	3	1	1	5	3
26	1	.	4	1	2	1	2	4
27	.	.	.	.	.	.	.	.
28	.	.	.	.	.	.	.	.
29	5	3	1	1	1	1	1	.
30	5	2	1	1	1	1	1	.
31	5	2	1	3	1	1	5	3
32	5	2	5	3	1	1	5	3
33	1	.	2	2	1	1	1	.
34	5	1	1	3	1	1	2	3
35	2	3	3	2	1	1	1	.
36	5	2	3	2	2	1	2	4
37	5	2	1	1	2	1	1	.
38	1	.	2	2	1	1	1	.
39	5	1	1	1	2	2	1	.
40	1	.	1	1	2	1	1	.
41	5	1	1	3	1	1	5	2
42	1	.	2	3	1	1	1	.
43	1	.	2	1	2	1	1	.
44	5	2	1	3	1	1	5	3
45	2	4	2	3	1	1	1	.
46	1	.	5	1	1	1	1	.
47	.	.	.	.	.	.	.	.
48	.	.	.	.	.	.	.	.
49	1	.	2	1	1	1	2	3
50	.	.	.	.	.	.	.	.
51	5	3	1	1	3	1	2	4
52	5	1	1	1	2	2	1	.
53	1	.	4	1	3	1	1	.
54	.	.	.	.	.	.	.	.
55	.	.	.	.	.	.	.	.
56	1	.	2	1	1	2	1	.
57	.	.	.	.	.	.	.	.
58	.	.	.	.	.	.	.	.
59	5	2	2	1	1	1	4	4
60	4	2	2	1	4	2	1	.
61	2	4	5	1	1	1	2	3
62	2	4	5	2	1	1	1	.
63	5	4	1	1	1	2	1	.
64	1	.	1	3	1	1	1	.
65	.	.	.	.	.	.	.	.
66	.	.	.	.	.	.	.	.
67	5	3	1	1	2	1	2	4
68	2	3	2	1	2	2	3	3

REC	AMBULATORY STATUS	AGE AMBULANT	ORTHOPAEDIC SURGERY	BLADDER TYPE	UPPER URINARY TRACT STATUS	UROLOGICAL SURGERY	BLADDER MANAGE-MENT	AGE CONTI-NENT
69	.	.	.	2	1	1	1	.
70	.	.	.	.	.	.	.	.
71	2	2	1	3	1	1	5	3
72	5	4	5	1	1	1	1	.
73	5	2	1	1	2	1	5	3
74	2	4	2	3	1	1	2	4
75	5	1	1	3	1	1	5	3
76	5	2	1	3	1	1	1	.
77	2	3	2	1	2	1	2	4
78	.	.	.	.	.	.	.	.
79	1	.	4	2	1	1	1	.
80	.	.	.	.	.	.	.	.
81	1	.	4	1	1	1	1	.
82	1	.	1	3	1	1	2	4
83	3	3	5	2	1	1	1	.
84	1	.	1	1	2	5	2	4
85	5	2	1	3	1	1	1	.
86	5	3	1	2	1	1	5	3
87	1	.	3	3	1	1	1	.
88	.	.	.	.	.	.	.	.
89	5	2	2	3	1	1	2	5
90	1	.	1	3	2	1	1	.
91	2	4	5	3	1	1	2	4
92	1	.	2	1	2	2	1	.
93	1	.	2	1	2	2	1	.
94	2	5	4	1	2	5	1	.
95	5	3	2	1	2	2	1	.
96	.	.	.	.	.	.	.	.
97	4	2	1	1	2	2	2	4
98	5	2	1	1	1	1	3	4
99	5	1	1	1	2	1	1	.
100	5	3	1	2	1	1	1	.
101	5	1	1	2	1	1	5	3
102	1	.	1	1	1	2	1	.
103	.	.	.	.	.	.	.	.
104	1	.	5	1	1	1	1	.
105	5	4	3	2	1	1	1	.
106	1	.	1	1	1	1	1	.
107	5	2	1	1	1	2	1	.
108	5	2	1	3	1	1	1	.
109	2	4	4	1	2	1	2	4
110	4	2	1	1	1	1	2	4
111	1	.	1	3	2	5	3	5
112	5	4	3	1	5	3	3	5
113	2	4	1	1	1	1	2	4
114	1	.	4	1	1	1	1	.
115	1	.	2	1	2	3	1	.
116	1	.	4	2	1	1	1	.
117	1	.	1	2	1	1	1	.
118	1	.	2	1	1	1	1	.

OUTCOME STUDY RESULTS

REC	DQ1	DQ2	DQ3	DQ4	GQA	GQB	GQC	GQD	GQE	GQF	GQ5	GQM
1	.	.	.	.	.	.	.	.	.	.	.	.
2	.	.	.	.	.	.	.	.	.	.	.	.
3	.	.	.	.	.	.	.	.	.	.	.	.
4	.	.	.	.	.	.	.	.	.	.	.	.
5	.	.	.	.	.	.	.	.	.	.	.	.
6	.	.	.	.	.	.	.	.	.	.	.	.
7	.	.	.	.	.	.	.	.	.	.	.	.
8	.	.	.	.	.	.	.	.	.	.	.	.
9	.	.	.	.	.	.	.	.	.	.	.	.
10	.	.	.	.	.	.	.	.	.	.	.	.
11	.	.	.	.	.	.	.	.	.	.	.	.
12	.	.	.	.	.	.	.	.	.	.	.	.
13	100	94	98	100	40	85	90	100	105	85	84	93
14	96	92	86	90	38	86	84	76	84	80	75	82
15	84	80	72	62	54	68	72	56	60	60	61	62
16	104	98	102	96	50	103	106	86	86	89	87	94
17	100	105	96	98	80	104	96	100	94	86	93	96
18	80	57	62	67	35	58	50	68	47	47	51	54
19	78	65	70	61	58	58	53	53	46	40	51	50
20	75	70	68	67	70	52	63	69	67	43	60	59
21	54	60	53	58	38	48	50	47	47	47	46	48
22	92	96	84	88	86	92	81	80	71	82	82	81
23	98	100	106	110	59	102	101	96	98	94	92	98
24	120	102	112	107	84	114	102	100	98	106	101	104
25	88	82	66	79	82	80	88	82	85	80	83	83
26	50	43	48	40	30	48	40	37	37	37	38	38
27	.	.	.	.	.	.	.	.	.	.	.	.
28	.	.	.	.	.	.	.	.	.	.	.	.
29	110	100	98	105	80	96	107	100	97	110	98	102
30	91	92	97	92	104	83	73	96	96	76	88	85
31	110	102	100	100	80	81	102	92	89	72	87	86
32	100	90	92	88	82	104	82	98	88	72	89	91
33	100	90	90	98	20	88	104	106	80	82	80	92
34	98	109	102	100	86	92	110	90	90	96	94	96
35	98	86	82	90	62	84	84	88	81	68	78	81
36	100	100	105	100	90	92	98	90	98	102	95	96
37	92	89	72	88	90	78	94	80	66	72	82	78
38	56	50	39	43	22	63	52	40	44	38	43	47
39	94	88	92	100	92	79	94	83	75	74	83	81
40	95	100	102	96	36	87	102	100	94	87	84	94
41	102	94	90	100	77	95	102	96	110	94	95	99
42	94	86	79	.	.	.	.	.	.	.	.	.
43	80	73	80	82	78	86	84	74	78	68	78	78
44	96	82	93	94	66	90	92	82	82	79	82	85
45	106	100	96	94	32	80	77	74	73	76	69	76
46	83	75	86	73	32	92	77	70	66	68	68	75
47	110	.	.	.	.	.	.	.	.	.	.	.
48	.	.	.	.	.	.	.	.	.	.	.	.
49	125	125	107	98	17	86	103	103	94	94	82	95
50	.	.	.	.	.	.	.	.	.	.	.	.
51	110	100	96	100	76	87	96	75	86	81	84	85
52	100	100	104	96	82	85	84	83	96	102	88	90
53	84	66	55	45	30	70	48	52	47	40	53	57
54	.	.	.	.	.	.	.	.	.	.	.	.
55	.	.	.	.	.	.	.	.	.	.	.	.
56	100	81	86	70	32	69	86	70	66	64	65	71
57	.	.	.	.	.	.	.	.	.	.	.	.
58	.	.	.	.	.	.	.	.	.	.	.	.
59	90	100	87	81	89	80	75	90	80	90	84	83
60	92	79	83	85	40	72	77	66	82	78	69	75
61	82	80	77	82	16	83	88	90	64	64	68	78
62	78	54	51	68	28	81	80	75	84	80	71	80
63	100	91	93	94	36	76	94	74	70	96	74	82
64	94	100	97	67	10	70	72	65	61	52	55	64
65	.	.	.	.	.	.	.	.	.	.	.	.
66	.	.	.	.	.	.	.	.	.	.	.	.
67	75	58	60	58	54	62	45	59	64	45	55	55
68	55	63	64	67	40	56	63	59	64	58	57	60

REC	DQ1	DQ2	DQ3	DQ4	GQA	GQB	GQC	GQD	GQE	GQF	GQ5	GQM
69	77	.	.	.	.	.	.	.	.	.	.	.
70	.	.	.	.	.	.	.	.	.	.	.	.
71	100	102	96	110	110	105	130	120	123	112	104	118
72	80	88	90	74	46	91	80	88	72	79	76	82
73	100	96	100	108	109	99	111	114	85	106	104	103
74	100	96	106	98	42	80	87	87	91	87	79	86
75	95	.	.	.	.	.	.	.	.	.	.	.
76	100	104	96	110	116	126	116	122	104	100	114	114
77	94	92	85	80	70	71	62	65	70	72	68	68
78	.	.	.	.	.	.	.	.	.	.	.	.
79	73	.	.	.	.	.	.	.	.	.	.	.
80	.	.	.	.	.	.	.	.	.	.	.	.
81	85	75	80	61	14	82	75	85	85	78	70	81
82	96	97	94	92	10	95	80	82	102	76	74	87
83	80	68	66	.	.	.	.	.	.	.	.	.
84	93	98	100	94	10	102	112	78	96	102	83	98
85	88	80	92	84	76	72	85	83	66	69	75	75
86	79	89	102	102	79	96	106	80	92	106	93	96
87	79	72	73	87	34	73	62	74	60	56	60	65
88	.	.	.	.	.	.	.	.	.	.	.	.
89	93	95	96	98	80	82	100	76	90	102	88	90
90	44	47	39	55	16	46	53	37	40	44	39	44
91	100	92	86	87	36	93	92	86	88	86	80	89
92	54	45	56	62	10	52	55	52	51	45	44	51
93	75	87	77	62	14	62	75	60	66	62	57	65
94	100	94	88	89	32	80	72	86	84	68	70	78
95	89	96	100	105	79	90	102	74	82	82	72	86
96	.	.	.	.	.	.	.	.	.	.	.	.
97	100	106	102	98	82	102	88	92	86	80	88	89
98	75	85	92	95	70	84	86	84	88	78	82	84
99	80	75	71	82	74	72	72	70	66	60	69	68
100	.	.	.	.	.	.	.	.	.	.	.	.
101	100	118	110	114	82	100	115	104	98	123	104	108
102	80	74	82	.	.	.	.	.	.	.	.	.
103	.	.	.	.	.	.	.	.	.	.	.	.
104	48	42	50	.	.	.	.	.	.	.	.	.
105	83	87	79	83	67	68	72	64	58	58	64	64
106	98	109	96	102	14	85	101	82	95	102	80	93
107	100	102	106	101	94	101	100	94	95	115	99	101
108	98	102	82	88	82	80	84	86	71	84	81	81
109	104	96	90	102	34	98	94	90	68	70	76	84
110	110	115	101	96	76	103	106	86	86	108	94	98
111	92	88	96	80	10	81	92	82	80	75	70	82
112	90	95	91	94	78	86	105	84	100	115	95	98
113	110	98	102	100	36	108	112	93	97	104	91	102
114	78	65	72	53	24	79	80	70	78	68	66	75
115	92	86	90	73	20	58	82	60	60	50	55	62
116	69	38	40	25	65	36	30	38	30	29	38	33
117	64	.	.	.	.	.	.	.	.	.	.	.
118	85	90	78	86	36	82	95	89	84	70	76	78

D.Q. - Developmental Quotient (DEI)
G.Q. - General Quotient (Griffiths)
1,2,3,4,5 - Age at assessment; M - Modified;
A,B,C,D,E,F - Griffiths Subscales

OUTCOME STUDY RESULTS

REC	OUTCOME1	OUTCOME2	OUTCOME3	OUTCOME4	OUTCOME5
1	.	.	.	.	.
2	.	.	.	.	.
3	.	.	.	.	.
4	.	.	.	.	.
5	.	.	.	.	.
6	.	.	.	.	.
7	.	.	.	.	.
8	.	.	.	.	.
9	.	.	.	.	.
10	.	.	.	.	.
11	.	.	.	.	.
12	.	.	.	.	.
13	10	4	4	4	4
14	10	10	4	4	4
15	10	10	4	5	5
16	10	10	4	4	4
17	4	4	1	1	1
18	10	11	11	11	11
19	5	6	5	5	5
20	10	10	8	2	2
21	11	11	11	11	11
22	10	10	4	1	1
23	10	10	4	4	4
24	10	10	4	1	1
25	10	4	1	1	1
26	11	12	12	9	9
27	.	.	.	.	.
28	.	.	.	.	.
29	10	10	4	4	4
30	10	4	4	4	4
31	10	4	1	1	1
32	10	4	1	1	1
33	10	10	10	10	10
34	4	4	1	1	1
35	10	10	4	4	4
36	10	4	4	1	1
37	10	4	4	4	5
38	11	11	12	12	12
39	4	4	4	4	4
40	10	10	10	10	10
41	4	1	1	1	1
42	10	10	10	.	.
43	10	10	10	10	11
44	10	4	1	1	1
45	10	10	10	4	5
46	10	10	10	10	10
47	10	.	.	.	.
48	.	.	.	.	.
49	10	10	7	7	7
50	.	.	.	.	.
51	10	10	4	1	1
52	4	4	4	4	4
53	10	11	11	5	5
54	.	.	.	.	.
55	.	.	.	.	.
56	10	10	10	10	11
57	.	.	.	.	.
58	.	.	.	.	.
59	10	4	4	1	1
60	10	4	4	4	4
61	10	10	7	1	1
62	10	11	11	5	4
63	10	10	10	4	4
64	10	10	10	10	11
65	.	.	.	.	.
66	.	.	.	.	.
67	10	11	5	2	2
68	11	11	2	2	2

REC	OUTCOME1	OUTCOME2	OUTCOME3	OUTCOME4	OUTCOME5
69	10	11	.	.	.
70	.	.	.	.	.
71	10	4	1	1	1
72	10	10	10	4	4
73	10	4	1	1	1
74	10	10	10	1	1
75	4	.	.	.	.
76	10	4	4	4	4
77	10	10	4	1	2
78	.	.	.	.	.
79	10	.	.	.	.
80	.	.	.	.	.
81	10	10	10	11	10
82	10	10	10	7	7
83	10	11	5	.	.
84	10	10	10	7	7
85	10	4	4	4	4
86	10	10	1	1	1
87	11	11	11	10	11
88	.	.	.	.	.
89	10	4	4	4	1
90	12	12	12	12	12
91	10	10	10	1	1
92	11	12	11	11	11
93	10	10	10	11	11
94	10	10	10	10	4
95	10	10	4	4	4
96	.	.	.	.	.
97	10	4	4	1	1
98	10	4	4	1	1
99	4	4	4	4	5
100	.	.	.	.	.
101	4	4	1	1	1
102	10	10	10	.	.
103	.	.	.	.	.
104	12	12	12	.	.
105	10	10	10	4	5
106	10	10	10	10	10
107	10	4	4	4	4
108	10	4	4	4	4
109	10	10	10	1	1
110	10	4	4	1	1
111	10	10	10	10	7
112	10	10	10	4	1
113	10	10	10	1	1
114	10	11	10	11	11
115	10	10	10	11	11
116	11	12	12	12	12
117	11	.	.	.	.
118	10	10	10	10	1

1,2,3,4,5 - Age in Years.

OUTCOME STUDY RESULTS

REC	S/M LEVEL @ BIRTH Grouped	S/M LEVEL @ 5 YEARS Grouped	S/M LEVEL @ BIRTH Coded	S/M LEVEL @ 5 YEARS Coded	S/MLEVEL @ 5 YEARS
1	4	.	12	.	.
2	6	.	16	.	.
3	4	.	12	.	.
4	2	.	9	.	.
5	3	.	11	.	.
6	3	.	11	.	.
7	4	.	12	.	.
8	3	.	10	.	.
9	2	.	9	.	.
10	4	.	12	.	.
11	1	.	7	.	.
12	4	.	12	.	.
13	4	3	12	11	L 4
14	4	3	12	11	L 4
15	5	5	14	13	S 1
16	5	4	13	12	L 5
17	6	6	16	15	S 3
18	2	3	9	11	L 4
19	6	6	16	15	S 3
20	5	3	13	11	L 4
21	3	3	10	11	L 4
22	5	4	13	12	L 5
23	5	5	13	13	S 1
24	5	4	14	12	L 5
25	6	5	16	13	S 1
26	3	3	10	10	L 3
27	3	.	11	.	.
28	4	.	12	.	.
29	4	5	12	14	S 2
30	5	5	14	13	S 1
31	5	5	14	13	S 1
32	5	5	14	14	S 2
33	5	3	13	11	L 4
34	6	5	15	14	S 2
35	4	4	12	12	L 5
36	5	5	14	13	S 1
37	6	6	16	15	S 3
38	2	3	9	10	L 3
39	6	6	16	16	S 4
40	4	3	12	11	L 4
41	6	6	15	16	S 4
42	3	3	10	10	L 3
43	3	3	10	11	L 4
44	5	5	13	13	S 1
45	3	3	11	11	L 4
46	3	3	11	10	L 3
47	6	6	17	15	S 3
48	2	.	9	.	.
49	1	1	7	6	T11
50	1	.	2	.	.
51	5	5	13	14	S 2
52	6	5	15	14	S 2
53	2	3	9	10	L 3
54	3	.	10	.	.
55	6	.	15	.	.
56	4	3	12	11	L 4
57	6	.	15	.	.
58	3	.	10	.	.
59	6	6	16	15	S 3
60	4	3	12	11	L 4
61	2	3	9	10	L 3
62	4	3	12	11	L 4
63	5	5	13	13	S 1
64	1	1	6	6	T11
65	3	.	11	.	.
66	4	.	12	.	.
67	6	5	15	14	S 2
68	5	5	14	13	S 1

REC	S/M LEVEL @ BIRTH Grouped	S/M LEVEL @ 5 YEARS Grouped	S/M LEVEL @ BIRTH Coded	S/M LEVEL @ 5 YEARS Coded	S/MLEVEL @ 5 YEARS
69	4	3	12	11	L 4
70	4	.	12	.	.
71	6	5	16	14	S 2
72	6	5	15	13	S 1
73	6	6	16	15	S 3
74	4	3	12	11	L 4
75	6	6	17	16	S 4
76	5	5	14	13	S 1
77	5	5	13	14	S 2
78	2	.	9	.	.
79	3	2	10	9	L 2
80	2	.	8	.	.
81	2	2	9	8	L 1
82	1	1	1	3	T 8
83	3	3	10	10	L 3
84	1	1	7	6	T11
85	5	5	14	14	S 2
86	5	5	14	13	S 1
87	3	3	11	11	L 4
88	1	.	3	.	.
89	4	5	12	13	S 1
90	3	2	10	8	L 1
91	3	3	10	11	L 4
92	3	2	10	9	L 2
93	3	2	10	9	L 2
94	3	3	10	11	L 4
95	5	5	14	13	S 1
96	2	.	8	.	.
97	6	6	15	16	S 4
98	6	5	15	13	S 1
99	6	6	16	16	S 4
100	4	.	12	.	.
101	6	6	16	17	S 5
102	3	2	11	9	L 2
103	5	.	14	.	.
104	4	3	12	11	L 4
105	5	4	13	12	L 5
106	2	2	9	8	L 1
107	6	6	17	17	S 5
108	5	5	13	13	S 1
109	3	3	10	10	L 3
110	3	4	11	12	L 5
111	2	2	8	8	L 1
112	4	4	12	12	L 5
113	4	3	12	11	L 4
114	2	3	8	10	L 3
115	4	3	12	10	L 3
116	4	3	12	11	L 4
117	2	2	9	8	L 1
118	3	3	11	10	L 3

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