ORTHOPAEDIC MANAGEMENT OF SPINA BIFIDA CYSTICA : THE CAPE TOWN EXPERIENCE

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CONTENTS

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PREAMBLE

This study outlines our experience with Spina Bifida Cystica (SBC) from an orthopaedic point of view at the Spinal Defects Clinic in the Red Cross Memorial Children's Hospital, Rondebosch, Cape Town.

An outline of the nature of the disease, the problems associated with it and our approach to their management will be highlighted in the introduction. This will be compared to other workers in the field abroad.

In the sections on clinical material and results the nature of the clinic and full documentation of the patients as far as possible is presented. This is followed by the discussion where our experience is contrasted with other workers in the field.

It is beyond the scope of this study to provide an in depth analysis of the results of surgical treatment of every deformity. Separate papers are planned to analyse our experience with deformities of the hip and foot.

The survival of by far the majority of these patients has been assured by the advent of antibiotics and advances in neurosurgery. In former years meningitis, hydrocephalus and urinary tract infections took a heavy toll during infancy and early childhood. Early surgical closure of

open/...

open lesions, ventriculo-peritoneal shunting procedures and the use of antibiotics have been factors contributing to survival.

The long-term survival of these patients, with their concomitant spine and lower limb deformities, has necessitated the incorporation of orthopaedic surgeons in their management.

It is advantageous for them to be treated in a combined specialist clinic rather than to be treated piecemeal in peripheral hospitals.

I N T R O D U C T I O N

Definition

The term spina bifida cystica (SBC) is synonymous with myelodysplasia and includes meningocele, myelomeningocele and myelocele. The condition is a neural tube defect which arises when there is failure of the embryological development of the spinal cord, the meninges and the posterior elements of the vertebrae. More detail will be presented later.

INCIDENCE

The worldwide incidence is influenced by these factors:

1. Racial factors. It is more common amongst caucasians than amongst coloured and blacks. In Cape Town the incidence of neural tube defects including anencephaly is 1 in 300 for whites, 1 in 1 200 for blacks and 1 in 1 250 for coloureds. (Cornell, Nelson, Beighton 1985) (Beighton 1977).

2. Regional variation. The incidence for caucasians in Australia, 0,95 per 1 000 live births (Simpson 1976) compared to 3 per 1 000 in England (Sharrard 1979) and our own ± 3 per 1 000 (Cornell 1983).

3. Sex. The condition is commoner amongst females (58%) in Menelaus' series (1980), this difference is statistically relevant. In this study there was a male preponderance in whites (56%) and coloureds (58%) and a female preponderance amongst blacks (66%).

GENETICS

The pattern of inheritance is multifactorial and recessive. The risk of having a second child with a neural tube defect is 4%. That for a 3rd child is 10% and the chances of a fourth, after having three, 25%. The risk of a person with SBC having a child with the disease is 4%.(Menelaus 1980).

Expectant mothers at risk should be investigated at between 15 and 17 weeks. Ultrasound scanning is carried out first and if this is negative amniocentesis is performed. The amniotic fluid is tested for the presence of alpha fetoprotein as ultrasound is not accurate in picking up SBC.

Using these two techniques all anencephalics and 80-90% of SBC are diagnosed antenatally. (Grace 1984).

EMBRYOLOGY

The formation of the neural tube commences at the 6 somite stage which is at 20 days post-fertilisation. The central nervous system (CNS) develops from the neural plate which is of ectodermal origin. The plate develops into a tube i.e. closure of the plate, which starts in the cervical region and proceeds cranially and caudally. The process is completed at the 27th day when the posterior neuropore closes (25 somite stage).

The diagram (Fig. 1) helps in the understanding of this process.

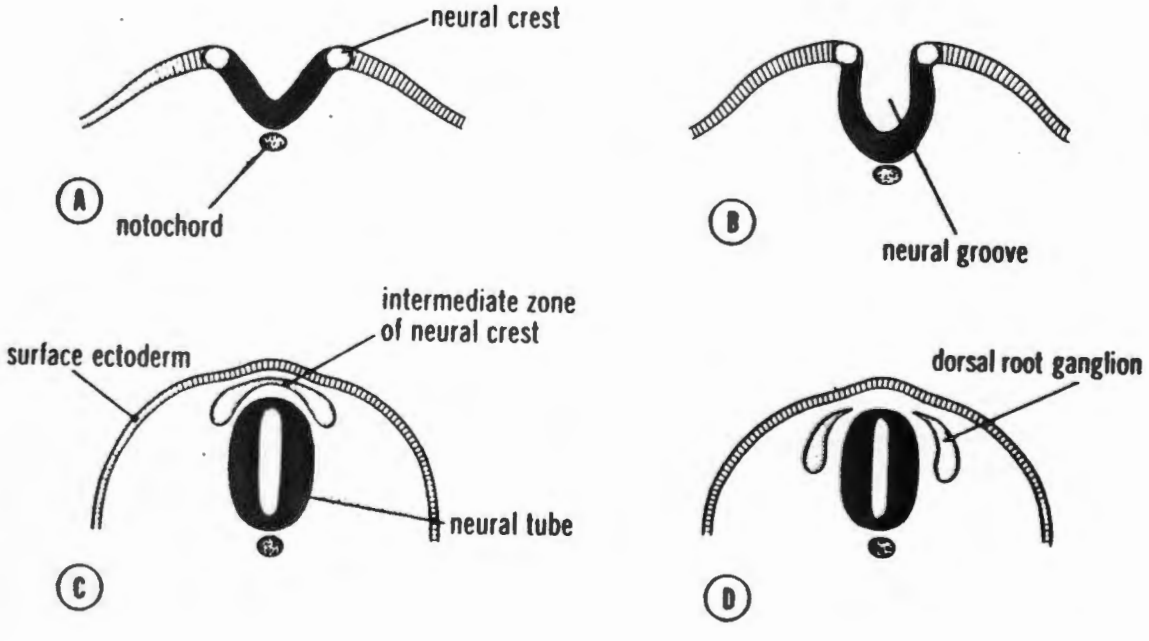


Fig 1. Simple line drawing to illustrate the closure of the neural plate to form the neural tube (Langman 1975).

The/...

The neuro-epithelium lining the neural tube then proliferates to form the various cell types that constitute the brain and the spinal cord.

Processes of the motor neurones grow out as axons of the peripheral nerves. Cells at the margins of the neural plate form a column on each side - the neural crest. This later forms sensory neurones of the dorsal root ganglion and their axons as well as the peripheral cells and processes of the autonomic nervous system. (See Fig. 1).

Much of the caudal extremity of the neural tube undergoes regression being represented eventually as the filum terminale which attaches to S3 vertebral body. At birth the cord terminates at L3 but gradually recedes up to L1 by adulthood.

The meninges are formed from loose mesenchyme surrounding the neural tube.

The vertebrae are formed from the somites, each of which contribute to two vertebrae. This results in the segmental blood supply being to two adjacent vertebrae.

The posterior chondral elements of the vertebrae develop at about 3 months. The severity of SBC depends on

the/...

the degree of failure in the development of these posterior elements. (Langman 1975).

Aetiology

The cause of the condition remains speculative. It seems that factors other than genetic are largely responsible. (Sharrard 1979). Studies of Monozygotic twins have shown concordance in 20%, for dizygotic twins the figure is 6%. This indicates an interplay of other factors such as environmental, social, maternal age and parity. (Sharrard 1979) (Laurence 1969).

Whatever the teratogenic factors are, they show their effect at a very early age, i.e. about 20 day-stage.

ANATOMY

The nature of the various types of SBC will be related here. Reference to figure 2 will aid understanding.

Spina Bifida Occulta (SBO)

This is a localised defect to one and occasionally more vertebral arches. It results from failure of fusion of the vertebral arches posteriorly. The spinal cord and meninges remain within the vertebral canal and are normal. This variant is present in 10% of the population at large and is usually discovered as an incidental finding. (Langman 1975). They are thus not patients seen at a Spinal Defects Clinic.

The presence of SBC may be indicated by a defect of the skin such as a dimple, patch of hair, pigmentation or a lipoma.

Spina Bifida Cystica

Meningocele. This is the least common variety where a fluid filled meningeal sac protrudes through the defect in the vertebral arches. The swelling is cystic and is covered by skin. The neural elements remain within the neural canal but may be functionally abnormal. The incidence is 0,76% in our series and 6% in Menelaus' series.

Myelomeningocele. The appearance is also of a cystic swelling covered by skin except here neural elements are present in the sac. The neural tube has closed normally. The most common lesion in our series 92%. In Menelaus' series it comprised 94%.

Myelocele

Also called myeloschisis or rachischisis. In our series it comprises 7%. Menelaus has a zero incidence in his series. It is the most severe form of the disease. Not only have the vertebral arches failed to form posteriorly but the neural plate is spread out over a cystic swelling of the meninges. If left alone it will eventually epithelialise from the edges. (Langman 1975) (See fig. 3).

Fig. 2/..

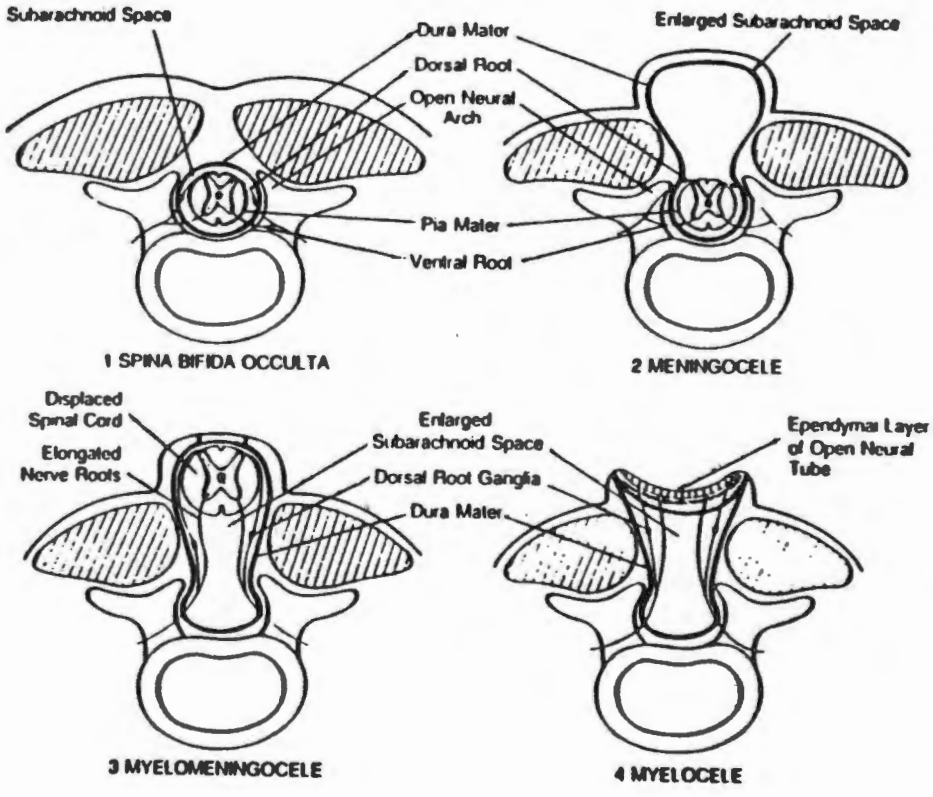


Fig. 2 Diagrams to illustrate the anatomy of the various types of spina bifida cystica. (Patten 1952).

The radiograph (Fig. 4) illustrates the appearance of the spine in SBC. Radiologically it is impossible to distinguish the 3 types of SBC. The SBO lesion is confined to 1 or 2 vertebrae and the defect is not nearly as widely spaced.

ASSOCIATED ABNORMALITY

SBC may be associated with the following congenital abnormalities:

1. Diastomatomyelia - the spinal cord is split by a bony bridge across the neural canal, this may in some cases be a fibrous septum.

2. Hydromyelia - the spinal cord is dilated by a cystic swelling of the central canal.

3. Associated skeletal defects:
   a) hemivertebra - which results from failure of complete formation of a vertebral body, this causes congenital kyphosis or scoliosis,
   b) failure of segmentation resulting in an unsegmented bar - again causing congenital kyphosis or scoliosis. (Moe et al 1978).

Fig. 3/...

Fig. 3 This photograph illustrates a myelocele which has epithelialised spontaneously.

Fig. 4/...

Fig. 4 Radiograph of the lumbar spine in SBC. Note the failure of formation of the posterior elements of L4 & 5 and the sacrum.

NEUROLOGY

The extent of the neurological lesion afflicting the legs is a combination of developmental dysplasia of the spinal cord and the effects of damage to the neural tissue by dessication, stretching, infection and compression by gliosis during healing - which can occur even after operative closure of the defect.

Stark & Baker (1967) describe 4 types of neurological lesion in SBC.

Type 1 The lesion affects the terminal part of the cord. Function is intact down to a certain level below which there is impairment of motor, sensory and reflex function. This may be partial or complete.

Type 11 Normal function of the cord to a certain level below which there is a gap with complete loss of motor, sensory and reflex function. Beyond the gap there is an isolated segment of functioning cord displaying reflex activity only. There is hyperreflexia, spasticity, no spontaneous movement and no sensation.

Type 111/...

Type 111 There is absence of function of a portion of the cord with some descending tracts passing through intact.

Type 1V No abnormality of cord function but abnormalities are present in the CNS above the lesion.

Types 1 and 11 are by far the commonest. Types 111 and 1V are rare. In their series Type 1 predominated in lumbosacral and sacral lesions. Type 11 were predominant in thoracic and thoracolumbar lesions and they were present equally in mid-lumbar lesions.

In our series we have not attempted to type the lesions in every case but have noticed a pattern of deformity in most cases which supports this theory, particularly types 1 and 11. Type 11 is considered to be present when, instead of flaccid paralysis below the level of the lesion, one encounters a rigid deformity present at birth.

The level of the lesion is dictated by the last functioning level, motor and sensory. All patients are labelled according to level in most centres.

Frequently/...

Frequently the level differs in the two legs by as much as two segments. We prefer to label ours as follows:

Thoracic lesions	-	paraplegic, no psoas function.
Upper lumbar	-	psoas and adductors (L1 - L4) functioning.
Mid lumbar	-	quadriceps i.e. active knee extension (L3 - 4).
Low lumbar	-	tibialis anterior (L4) and dorsiflexors (L4 - L5) functioning.
Sacral	-	gluteus maximus and plantar flexion (S1 - 3) functioning.

The vast majority of cases (95%) are incontinent of faeces and urine.

Menelaus' (1980) system of levels differs slightly from ours in that he has high and low thoracic level. High lumbar is the same but he includes mid-lumbar with low lumbar and upper sacral. His lower sacral group have no paralysis of the leg whatever but do have pelvic floor paralysis.

Sensory/...

Sensory loss in areas of the body where bony prominences are subcutaneous can result in the development of pressure sores. In thoracic lesions the sacrum and greater trochanter are particularly at risk whereas in mid and lower lumbar lesions the heels are.

Hydrocephalus

In this series the incidence of ventriculo-peritoneal shunting is 80%.

Intelligence

80% of these patients have normal intelligence. One patient who has mild hydrocephalus and a ventriculo-peritoneal shunt has an I.Q. of 155.

FUNCTION

Mode of ambulation is described in the following way:

1. Community walker - walks with ease for any distance.

2. Household walker or functional walker - walks about at home only, usually with the aid of appliances and crutches.

3. Non-functional walker - can walk only in calipers and crutches with the help of someone else. In this series patients in this category are regarded as non-walkers as they usually use wheelchairs.

4. Non-walker - wheelchair user.

DEFORMITY

The causes of deformity are threefold:

a) Muscle imbalance

b) Habitually assumed posture dictated by gravity.

c) Coexistent congenital deformity.

a) Muscle imbalance. Unapposed muscle groups will act producing a position which eventually becomes fixed. This is very pronounced if the unapposed muscle group is spastic. Rigid deformities present at birth are often caused by this.

Rigidity of the deformity develops because fasciae, capsule, ligaments and the muscles themselves become shortened. The deformity is worsened by growth because of the tethering effect of these tissues. Bony deformity occurs only after the deformity has been present for a long time.

The deformity starts in utero and continues during postnatal growth. Deformities present at birth tend to be more fixed than those occurring later.

b)/...

b) Habitually assumed posture. Tends to become fixed as the soft tissue structures on the concave side become shortened. Muscle imbalance is partly responsible for this but is assisted by gravity. The flail joint takes up a position dictated by gravity and eventually becomes fixed. This applies to the paralysed spine too.

After birth the flail leg takes up a position of abduction flexion and external rotation at the hip with flexion at the knee and equinus feet. (Fig. 5).

c) Coexistent congenital deformity. Congenital dislocation of the hip will contribute towards external rotation and adduction contracture. Hemivertebrae will cause scoliosis or kyphosis.

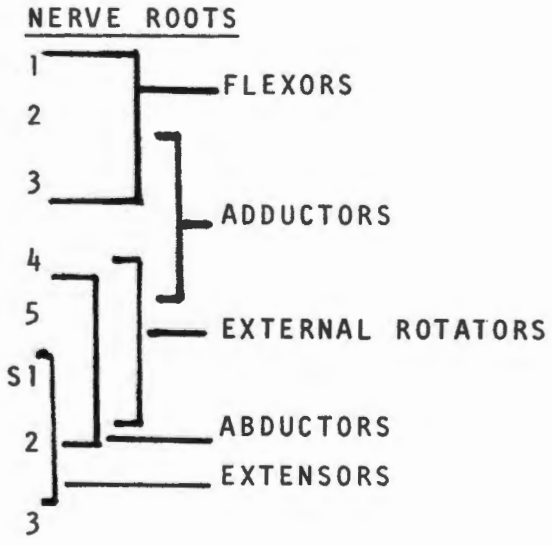
Deformity at the joints of the lower limb and spine will now be discussed in more detail.

Hip Deformity

The presence of hip deformity and the nature thereof is dependent on the level of the lesion.

In thoracic and L1 lesions the position of the leg is dictated by gravity i.e. flexion-abduction and external rotation. Dislocation is not a problem in these patients. Weak psoas action aids maintenance of this position. (Fig. 5).

In lesions with levels more distal to this, progressing down the cord, the adductors and flexors increase in power producing a position of flexion and adduction which predisposes to dislocation. Subluxation is more likely in L2 - L3 and dislocation in L3 - L4 lesions. Subluxation and dislocation are thus the most common in high and mid-lumbar lesions - our findings support this. Fig. 6 is a radiograph of a dislocated hip.



The above diagram illustrates the segmental innervation of the hip musculature.

Fig. 5/...

Fig. 5 Characteristic posture of a child with a thoracic or L1 lesion. Note flexion abduction and external rotation at the hip, flexion of the knee and equinus feet.

Abductor/...

Abductor activity begins at L4 and reaches full strength at S1.

Flexion deformity will occur down to S1 as gluteus maximus (S1 - S2) has not reached full power. The lateral hamstrings (L5 - S1) act as hip extensors as well and thus represent the first opposition to flexor activity starting in L5 lesions.

Hip deformity may also be caused by lumbar scoliosis which results in pelvic obliquity. Abduction deformity of one hip and adduction deformity of the other results. Dislocation of the adducted side develops if pelvic obliquity is not corrected.

Knee Deformity

Deformity here is less likely than at the hip or foot. Flexion, hyperextension (recurvatum) or valgus deformities may occur.

Flexion deformities occur in flail limbs and also in high lumbar lesions which have paralysed quadriceps muscles in the presence of knee flexors.

Recurvatum deformity may develop in L3 - L4 lesions where the quadriceps are strong and the hamstrings weak. Sartorius and gracilis become displaced anterior to the knee acting as aberrant extensors. (See Fig. 7).

Fig. 6/...

Fig. 6 A radiograph of a dislocated hip.

Fig. 7 This child has bilateral genu recurvatum and equinovarus feet.

This/...

This deformity is however rare considering only 6 out of 40 patients, with mid-lumbar lesions, had it.

Valgus deformity may develop in a flail leg or in a leg that has a spastic biceps femoris.

Foot Deformity

Deformities of the feet are present in over half of all SBC cases. (Menelaus 1980). In our series the incidence is 68%. The whole spectrum of foot deformities may occur. Calcaneovalgus and equinovarus are the most common. The more common varieties will be discussed.

Calcaneus Deformity

Occurs in L4 and L5 lesions where dorsiflexors are active and calf paralysis exists. Usually varus or valgus occurs concomitantly. Calcaneovalgus was our most common foot deformity. Theoretically tibialis anterior (L4) should cause a calcaneovarus deformity. This does occur but it seems that an alteration in the line of action of tibialis anterior occurs in these patients causing calcaneovalgus deformity. (See Fig. 8).

Calcaneus deformity is particularly disabling in SBC as the heel is also anaesthetic in L4 - L5 lesions and tends to ulcerate. A prominent heel is difficult to accommodate in a shoe and is difficult to walk on. (See Fig. 9).

Fig. 8/...

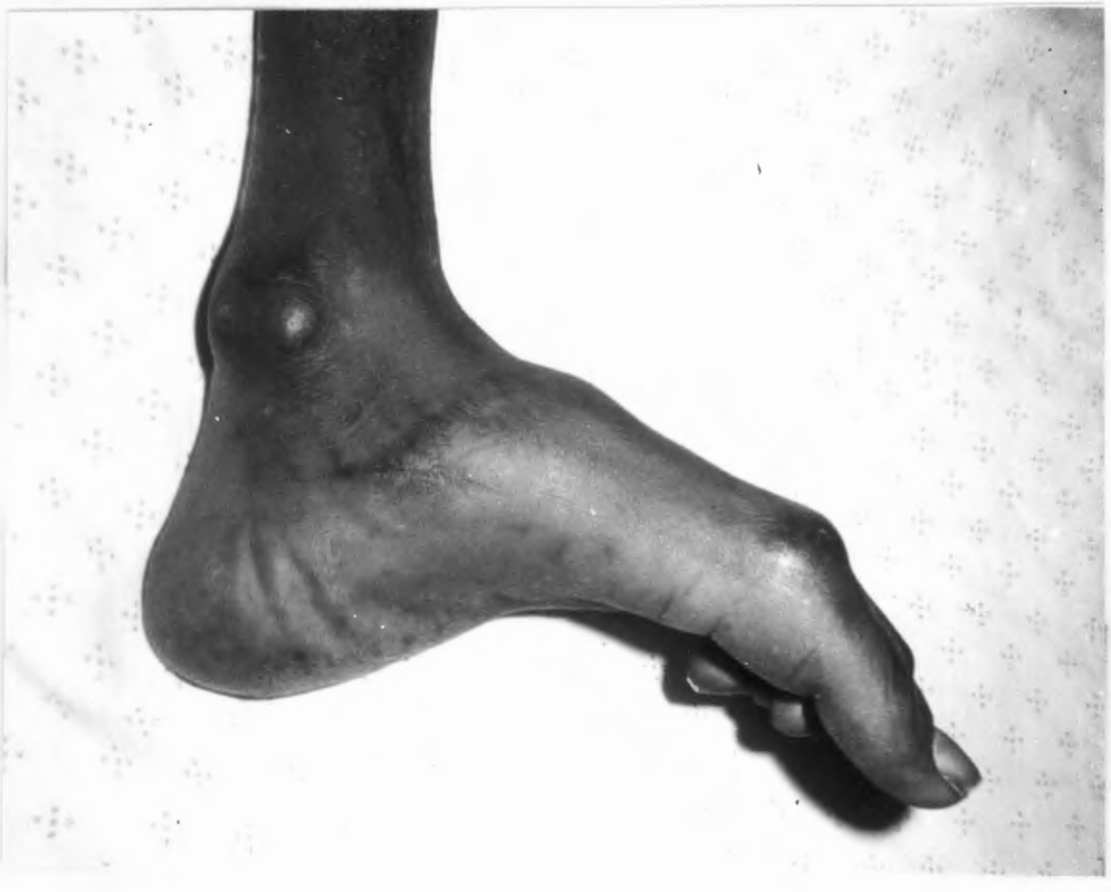


Fig. 8 Calcaneus foot deformity in an older child.

Fig. 9/...

Fig. 9 Heel ulceration in a child with calcaneus left foot.

Equinus/...

Equinus

The flail foot tends to gravitate into equinus. Equinovarus results from muscle imbalance usually, in a Type 11 (Stark & Baker) lesion, where the calf is spastic and overrides the dorsiflexors. This condition is usually present at birth tending to be rigid. (See Fig. 10).

Vertical Talus

Occurs in 1 - 10% of feet in different series, 1,8% in this series. It is thought to occur when the intrinsic muscles are paralysed in the presence of strong or even spastic dorsiflexors and plantar flexors. There is equinus of the hindfoot and dorsiflexion of the forefoot with dislocation at the mid-tarsal joint. The deformity so produced is called the 'rocker-bottom' foot. (See Fig. 11).

Clawing of the Toes and Cavovarus

Common in low sacral lesions apparently due to paralysed or weak intrinsics. The toe extrinsics, dorsiflexors and plantar flexors are functioning normally and are not spastic. Surgery may occasionally be necessary.

Rotational Deformities of the Tibia

May occur i.e. external rotation or internal rotation (internal tibial torsion). The former usually occurs in valgus and the latter in varus foot deformity.

Fig. 10/...

Fig. 10 Equinovarus feet in child. Tibial internal rotation is also evident.

Fig. 11 Radiograph of a vertical talus.

The/...

The Spine

Deformities encountered are kyphosis, scoliosis and lordosis. Aetiologically two groups are seen, congenital and paralytic.

Congenital

In addition to the usual dysgenesis of the posterior elements, other congenital malformations may be present. The defect may be a lack of formation or segmentation resulting in hemivertebra or unsegmented bars respectively. The incidence is said to be around 30%.

Developmental or Paralytic

Straight spines at birth, later developing deformity by way of a paralytic curve. 50% of SBC develop this deformity. (McLaurin 1976).

Discussion on the different deformities of the spine will now follow:

Kyphosis

The incidence is 5% of all SBC (Sharrard 1979), a figure which is surprisingly low considering the absence of posterior elements and the perverted action of the erector spinae and quadratus lumborum muscles which become flexors. Our figure is 4% overall.

The incidence is higher in thoracic lesions i.e. 10% (Sharrard 1979). In this series 33% had kyphosis.

Lordosis

This deformity occurs as a compensatory measure to ensure spinal balance. Flexion deformity of the hips produces a lumbar lordosis as does thoracic kyphosis.

Scoliosis

The commonest cause is paralytic in SBC, usually a long 'C'-shaped curve from mid-thoracic to the sacrum. Pelvic obliquity frequently develops presenting a problem with sitting and the development of pressure sores on the side of the convexity. The curve may be double with the lumbar element being lordotic.

Congenital scoliosis may occur anywhere in the spine and tends to be progressive and fixed. The two types may be present concurrently.

Incidence of Scoliosis (Menelaus 1980)

Thoracic Lesions

	Cape Town	Menelaus
Birth	0	1% - have curves > 30 degrees
4 years	16%	17% - have curves > 30 degrees
20 years	–	88% - have curves > 30 degrees

High/...

High lumbar lesions

	Cape Town	Menelaus
Less than 2 years	0	0 - have curves > 30 degrees
4 years	16%	14% - have curves > 30 degrees
20 years	–	81% - have curves > 30 degrees

Low lumbar lesions

	Cape Town	Menelaus
Birth	1%	3% - have curves > 30 degrees
4 years	0	18% - have curves > 30 degrees
20 years	–	23% - have curves > 30 degrees

Sacral lesions

	Cape Town	Menelaus
1 year	2%	3% - have curves > 30 degrees
20 years	–	9% - have curves > 30 degrees

The above comparison was obtained by temporarily adopting Menelaus' system of levelling. No patients in this series have reached 20 years, the oldest being 18 years (only 6 are over 15 years of age).

If curves greater than 10 degrees are considered the overall incidence is 11% and for curves over 30 degrees the incidence is 2,6%.

Banta (1976) had an incidence of scoliosis of 16% in patients under 5 years of age. At 15 years of age the incidence was 50%. Of the 6 patients in our series who have reached 15 years of age none have scoliosis with curves over 30 degrees and 2 have curves over 20 degrees. (33% incidence).

Fractures/...

Fractures

Paralysis causes osteoporosis which predisposes to fractures. Immobilisation in plaster casts aggravates this problem. It is thus not surprising that the incidence of fractures is as high as 18%. (Menelaus 1980). In this series the incidence of 2% is very low by comparison.

Radiographs of fractures in SBC (Fig. 12) show exuberant callus which can easily lead to a misdiagnosis of an osteosarcoma. The excessive callus is because of anaesthesia allowing movement and late diagnosis.

Fig. 12 Radiograph of a fracture in SBC showing exuberant callus which resembles an osteosarcoma.

Principles/...

PRINCIPLES OF MANAGEMENT

Newborn babies with SBC are sent to the neurosurgeons at Red Cross War Memorial Children's Hospital for closure of the lesion and ventriculo-peritoneal shunting if necessary. Their policy is to treat all comers regardless of the level or of the severity. The results of this approach have been shown (Black 1979) (McLone 1985) to differ little from selective treatment of cases (Lorber 1972) and certainly avoids the consideration of passive euthanasia with its concomitant emotional and medico-legal implications.

Infants receiving in-patient neurosurgical care are sometimes referred to the orthopaedic clinic if obvious deformities exist - usually club feet or dislocated hips. Serial plaster cast treatment is commenced in these patients if necessary. It is more usual for orthopaedic assessment of new cases to take place at the first clinic visit at 1 - 2 months of age.

At the first visit we assess the patient in terms of:-

a) level of lesion,
b) muscle power chart (as far as is possible in a baby),
c) deformity,
d) radiology of the spine and hips.

An/...

An attempt is made at predicting the ultimate function of the patient which allows realistic planning of surgery. This assessment is continued at subsequent visits, usually 3-monthly. Definite goals in terms of function are established at an early stage. The parents are informed at the outset of what we are doing and what they can expect in the future.

Early correction of deformity is achieved by soft tissue release alone whereas later on bony procedures may become necessary. The object is to straighten the limb so that ambulation is possible with or without the use of calipers, regardless of the level.

This approach is not applied in cases of mental retardation or spasticity. Deformity in these cases is corrected later, and only if wheelchair sitting or hygiene are compromised. Surgical management of specific deformities will be described later.

To/...

To achieve ambulation in patients with high lesions, external support by above knee calipers and crutches (Fig. 13) and the use of a 'swing-through' gait, is the only way these children can walk. This method is adequate until adolescence when the power-weight ratio of the patient changes and a wheelchair is usually resorted to, being far less tiring. (Menelaus 1983) (Asher 1984).

It is our practice to get the patients with high lesions into a standing frame at about 18 months. The object is to develop the desire to ambulate as the brain is stimulated by the new proprioceptive and other sensory input. (Menelaus 1980).

The use of physiotherapy is somewhat limited by distance as many of these patients are from up-country. When patients show no desire to walk, the effort is abandoned.

Below knee calipers (Fig. 14) or ankle foot orthoses (AFO) (Fig.15) are used in patients who have knee control but require help in maintaining a plantigrade foot. To counter the tendency to pressure sores in anaesthetic feet, fur-lined boots (Fig.16) are used. Well made AFO's do not cause pressure sores and, if possible, should be used as they are lighter and more acceptable than calipers.

Fig. 13/...

Fig. 13 This child has a high lumbar lesion but is able to ambulate with long-leg calipers and crutches using a swing-through gait.

Fig. 14/...

Fig. 14 Below knee calipers have been used extensively until now but are slowly being replaced where possible, with ankle-foot orthoses (AFO).

Fig. 15/...

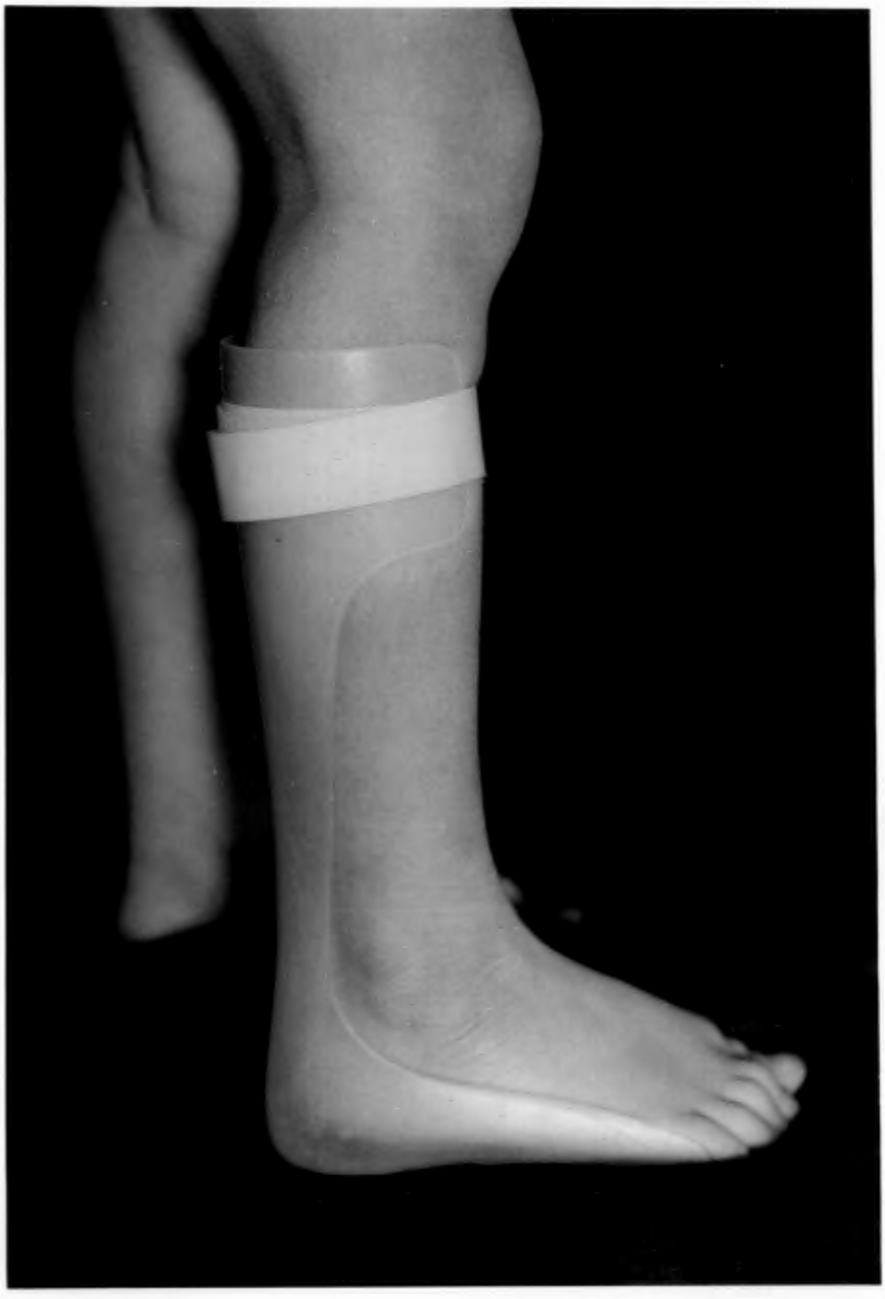


Fig. 15 Photograph of an AFO made of polypropylene. This substance becomes malleable when heated and thus lends itself to being used for bracing.

Fig. 16/...

Fig. 16 An example of the type of fur-lined boot made at the Red Cross Hospital Workshop, which is used on all patients whether calipers or AFO's are worn.

Surgical/...

SURGICAL MANAGEMENT OF THE SPINE

The patients are screened clinically for spine deformity at each visit. For radiological assessment we make use of the intravenous pyelograms (IVP) ordered by the urologist usually at 6-monthly intervals. If necessary, X-rays of the spine are requested.

Spinal deformities that progress to the stage where treatment is required are referred to spinal surgeons specifically interested in this difficult problem.

Scoliosis

Scoliotic curves are usually paralytic or combinations of paralytic and congenital. Pure congenital scoliosis, if progressive, requires posterior fusion as bracing is of little value. (Moe et al 1978). Mixed curves and paralytic curves do respond to bracing which is applied in progressive curves over ± 30 degrees. The spinal unit make use of polypropylene tailor-made thoraco-lumbo-sacral orthoses (TLSO) (Fig. 17).

Fig. 17/...

Fig. 17 The type of TLSO constructed by the Red Cross Hospital Workshop.

Surgery/...

Surgery is considered only if progression occurs in spite of bracing or if the patient is unbraceable i.e. curves greater than ± 60 degrees, or if pressure sores occur.

Two-stage surgery is performed if there is a lordotic lumbar element to the curve. The first stage is an anterior fusion of the lumbar spine T12 to L5 by excising the discs and applying Dwyer instrumentation. At least two weeks later the spine is fused posteriorly to include the whole extent of the curve or curves. Stability and correction is provided by using Harrington distraction rods and modified Luque segmental wiring of the spinous processes to the rod.

Posterior fusion alone is used when the curve has a kyphotic element to it i.e. kyphoscoliosis instead of lordoscoliosis. This is because the excision of discs and Dwyer instrumentation will aggravate the kyphosis.

The object of this form of treatment is to straighten the spine and level the pelvis thus avoiding ischial pressure sores on the convex side of the curve. It also frees both arms for use other than body support which is especially important in SBC cases. (Menelaus 1980) (Turner 1985). The approach used by other authors is

the/...

same as ours and has its origins in Twin cities (Moe, Winter, Bradford and Lonstein) and in Toronto, (Hall).

Kyphosis

This deformity is usually operated on when the severity warrants it: bracing is ineffective (Menelaus 1980). The main indication for surgery is recurrent ulceration over the apex of the kyphosis. Another relative indication is to allow access to the abdomen for urinary bypass procedures. (Menelaus 1980). Sharrard mentions two additional indications: increasing kyphosis and neurological involvement.

The basic procedure common to all is excision of the apical vertebra and some of the vertebrae above and below the apex and then fusing these two together. The means of fixation varies. Sharrard uses staples, Menelaus used threaded 'K' wires in children under 5 years, over this age he favours Harrington compression apparatus. He also fuses the vertebrae for two levels above and below, both anteriorly and posteriorly.

No patient in our series has required surgery. The urologists no longer use ileal conduits, thus removing one indication. None of the 9 cases with kyphosis have recurrent ulceration.

MANAGEMENT OF HIP DEFORMITY

In patients who have walking potential it is essential to treat deformity of the hip, as the presence of deformity severely affects function. (Asher 1983). Hip dislocation without deformity does not preclude walking. (Riggins 1983) (Schaffer 1983).

Flexion Adduction Deformity

This is actively treated early on as it predisposes to dislocation if allowed to persist. We have been satisfied with 14 patients in whom surgical release of the adductors and iliopsoas was done bilaterally at one sitting. Post-operatively the correction is maintained in well padded broomstick plaster casts for six weeks. (Fig. 18). Other authors treat this deformity in the same way. (Menelaus 1980) (Schaffer 1983).

Fig. 18/...

Fig. 18 An example of a child in so-called broomstick plaster casts to hold the hips abducted following iliopsoas and adductor release.

Sharrard/...

Sharrard proceeds to postero-lateral transfer of the iliopsoas (Sharrard 1964) if the hip continues to sublux in spite of adductor release. The procedure involves removing iliopsoas from the lesser trochanter and transferring it through the wing of the ilium to the postero-lateral aspect of the greater trochanter. Menelaus (1980) has done many of these transfers but appears to be less enthusiastic about it. (Stillwell and Menelaus 1984). He has shown the transferred muscle to be inactive and the only benefit over simple iliopsoas excision is that it has a tenodesis effect while weight-bearing.

Six Sharrard procedures have been done in our unit but the procedure was abandoned in 1981 in favour of simple flexion-adduction release. It seems unnecessary to perform major surgery when a simple procedure will achieve the same result.

Flexion Deformity

Pure flexion deformity in excess of 20-30 degrees after the first year of life is surgically corrected by releasing the hip flexors and, if necessary, the anterior joint capsule. Correction is maintained post-operatively by lying the patient supine on a convex bed with an above knee broomstick plaster in abduction. (Fig. 18). Eighteen such procedures have been performed in our unit.

Flexion/...

Flexion Abduction External Rotation Contracture

This is the position a paralysed limb takes up as dictated by gravity. Confined in our series to thoracic (8 cases) and high lumbar lesions (1 case). The position becomes fixed with time and may require surgical release. The external rotators and posterior capsule are released and in addition the iliopsoas and other flexors - if necessary. Only one patient in our series required this release.

Hip Dislocation

The Pavlik harness is used in the infant for at least three months. The use of the harness is limited to those without significant adduction deformities. Some authors (Menelaus 1980, Sharrard 1974) condemn its use, saying that a flexion abduction external rotation deformity is encouraged.

In those where an adduction deformity precludes the use of the harness we perform a surgical release of adductors and flexors. We usually find that the hip reduces very easily as no limbus is present as in congenital dislocation of the hip.

Open reduction of the hip becomes necessary on the rare occasions when reduction is not attained conservatively. This is particularly indicated when the dislocation is unilateral and there is grade 4/5 quadriceps power.

Other/...

Other factors are a good potential for life and less than 5 years of age. Bilateral dislocation makes the decision difficult particularly since it has been shown that function is not impaired by dislocation in these cases. (Riggins 1985). We have tended to be very conservative in this regard - 4 open reductions in 3 patients (1 bilateral) out of 42 dislocations.

If an open reduction is performed, the adductors and psoas must also be released to avoid recurrence. Following this procedure a varus derotation osteotomy is performed. (See Fig. 19). Sharrard (1974) dislikes this procedure as he feels that muscle imbalance has not been dealt with and that his iliopsoas transfer addresses the problem more adequately. Clearly a controlled study is required to settle the dispute.

Innominate osteotomy is necessary in some cases where the acetabulum is very dysplastic and subluxation is present. The indication for surgery in this situation is unclear as function is not altered by subluxation in SBC. In our series there were 25 subluxed hips in 18 cases of which 58% were able to walk. Chiari osteotomy has been recommended in the older child and Pemberton osteotomy in the younger child. (Menelaus 1980). We have performed 3 Chiari osteotomies.

Fig. 19/...

Fig. 19 Varus derotation osteotomy of the left proximal femur which has successfully contained the hip.

MANAGEMENT OF KNEE DEFORMITY

Flexion Deformity

Flexion deformity over 20 degrees should be corrected as function is impaired. If hip flexion deformity coexists then it is advisable to correct the hip first after which the knee is treated if deformity is still present. It is better to do this under one anaesthetic and thus diminish the total time spent in Plaster of Paris. (Menelaus 1980). Prolonged immobilisation tends to aggravate the osteoporosis already present.

Surgical correction involves tenotomy of hamstrings at the knee. The posterior knee capsule and cruciate ligaments may need to be divided in addition. This procedure was successful in 7 patients (6 bilateral) i.e. ambulation possible post-operatively whereas pre-operatively they were not walking.

Many flexion deformities, present in the first year of life, correct spontaneously. (Schaffer 1983, Menelaus 1980). Surgical release should therefore be postponed until later. However, if hip surgery is being performed it is wise to correct a significant knee deformity at the same time.

In/...

In children older than 10 years, corrective supra-condylar osteotomy becomes necessary. (Menelaus 1980 and Schaffer 1983). If done under this age, the chances of recurrence are high.

Recurvatum

Overactive or spastic quadriceps muscles may be surgically elongated if the child has good potential as a walker. (Menelaus 1980). Our approach is to leave the deformity as it lends itself to wearing a long-leg caliper.

If the child is a wheelchair user then tenotomy of the patellar tendon is practised.

MANAGEMENT OF FOOT DEFORMITY

Equinovarus Deformity

The ideal approach is corrective plaster casts and surgery if necessary. The more rigid deformity improves after a subcutaneous tenotomy of the calcaneal tendon which is performed together with serial plaster casts. If this fails to achieve adequate correction then a postero-medial release is indicated. (Menelaus 1980, Sharrard 1979).

It is, however, impractical to get the majority of our patients back weekly for plaster changes. We have thus adopted a no treatment regime in many cases, performing a postero-medial release near to walking age. Retention plasters are used for six weeks and then Denis Brown corrective boots until ambulation is well on its way. Recurrence of the deformity is common (Menelaus 30%), and 29% in our series (11 out of 37 operations).

Recurrences are treated by repeat postero-medial releases. Menelaus (1980) does the same but Sharrard (1979) advocates talectomy.

Resistant cases over 10 years of age are treated by doing a triple arthrodesis.

Equinus/...

Equinus Deformity

This is effectively treated by doing subcutaneous tenotomy of the calcaneal tendon. Usually no anaesthetic is necessary. Post-operatively a well padded cast is used for three weeks, after which the patient is put back into his AFO or caliper with backstop, if one is required.

Severe equinus deformities require a more complete posterior release. Other authors have the same approach. (Schaffer 1983, Menelaus 1980).

Cavovarus Deformity of the Heel

The standard procedure for this deformity is release of plantar fascia (Steindler strip) from the calcaneum and a Dwyer lateral closing wedge osteotomy of the calcaneum. This is best done after 3 years of age. We have done 5 of these procedures.

Claw Toes

This deformity has not been troublesome in our experience except in 3 big toes (1 bilateral). These were treated by doing Jones' procedure for the big toe. This involves transferring the extensor hallucis longus to the neck of the 1st metatarsal and tenodesing the flexor hallucis longus to the middle of the proximal phalanx. Sharrard has a similar approach. (Sharrard 1979).

Calcaneus/...

Calcaneus Deformity

We have had success with the Peabody procedure which involves transfer of the tibialis anterior to the heel through the interosseous membrane. This is done when the patient starts walking. So far 36 such procedures have been performed. The transfer has to be performed in addition to soft tissue release for contractures i.e. anterior ankle or lateral ankle in calcaneo-valgus deformity. The peronei are lengthened and not transferred to the heel as recommended by Sharrard. (Sharrard 1968).

In older children (4-12 years) when a pistol grip heel has developed, a calcaneal osteotomy is performed. This is posterior displacement of the calcaneal tuberosity after the body of the calcaneus has been divided.

In patients over 12 years of age a triple arthrodesis using the Elmslie technique is employed. (Cholmeley).

Vertical Talus

Rocker-bottom foot deformity is usually managed in two stages: the first is an anterior release of all tight soft tissues and reduction of the mid-tarsal joint, which is held with two 'K' wires. The tibialis anterior tendon is transferred to the neck of the talus. The second stage is performed three weeks later

when a posterior release is performed. Menelaus does it all in one sitting.

The operation is done at 3 - 6 months of age. We have done 7 of these operations in 4 patients. So far none have recurred.

CLINICAL MATERIAL

This study was done at the Spinal Defects Clinic which is held in the Physiotherapy Department at the Red Cross War Memorial Children's Hospital. It is held every Wednesday afternoon and has been in existence since 1972.

In attendance at this clinic are the following:-

- Orthopaedic Surgeon.
- Urologist.
- Neurosurgeon.
- Physician specialised in developmental aspects of childhood.
- Physiotherapist.
- Social Worker.
- Genetics Councillor.
- Stomatherapist.

The patients circulate from one specialist to the next during routine visits which are 3 to 6 monthly. Problems in a particular area may necessitate weekly visits to the specialist involved, the others only being seen when the necessary time has elapsed. In-patient treatment, in all specialities with the exception of orthopaedics, is carried out in the Red Cross War Memorial Children's Hospital. The lack of orthopaedic bed space there necessitates admission

to/...

to either Maitland Cottage Home or Princess Alice Orthopaedic Hospital. The latter is used for major surgery because of the better after-care facilities.

Patients are referred to this clinic from the entire Cape Province, Ciskei and Transkei, a total population of approximately 12 million.

This study has been confined to SBC thus simplifying interpretation and allowing comparison with other publications. They are also by far the most numerous. Other conditions seen are lipomas of the cord, sacral agenesis, diastematomyelia, transverse myelitis, post-traumatic paraplegia and a very small number of TB spine, though this condition is not usually treated there by us.

The study group consists of 188 cases with an age range of one month to 18 years with a mean of 6,5 years. Many cases seen at the clinic since its inception in 1972 had to be excluded because of no follow-up in the study period. The study period was 1st June 1983 to 31st October 1983 then again 1st August 1985 to 31st October 1985.

The age limit at the Red Cross War Memorial Children's Hospital is 13 years. This rule fortunately is relaxed in SBC cases who attend well into their teens if

necessary. Eventually they are transferred to Groote Schuur Hospital or Conradie Hospital where they are managed piecemeal by the various specialities whenever problems arise.

The orthopaedic team there at present is Dr Lewis Sparks, consultant orthopaedic surgeon and the registrar working at Maitland Cottage Home.

The patients were assessed for deformity, neurological level, muscle charting and function. Radiographs of the hip and spine were examined to exclude deformity.

RESULTS/...

RESULTS

Race and Sex Incidence

The study group consists of 101 males and 87 females. (Table 1). Male preponderance exists in our white and coloured race groups but is reversed in the black group.

TABLE 1

	WHITE	COLOURED	BLACK	TOTAL
Male	27 (56%)	64 (58%)	10 (34%)	101 (53%)
Female	21 (44%)	47 (42%)	19 (66%)	87 (47%)
Total	48 (26%)	111 (59%)	29 (15%)	188

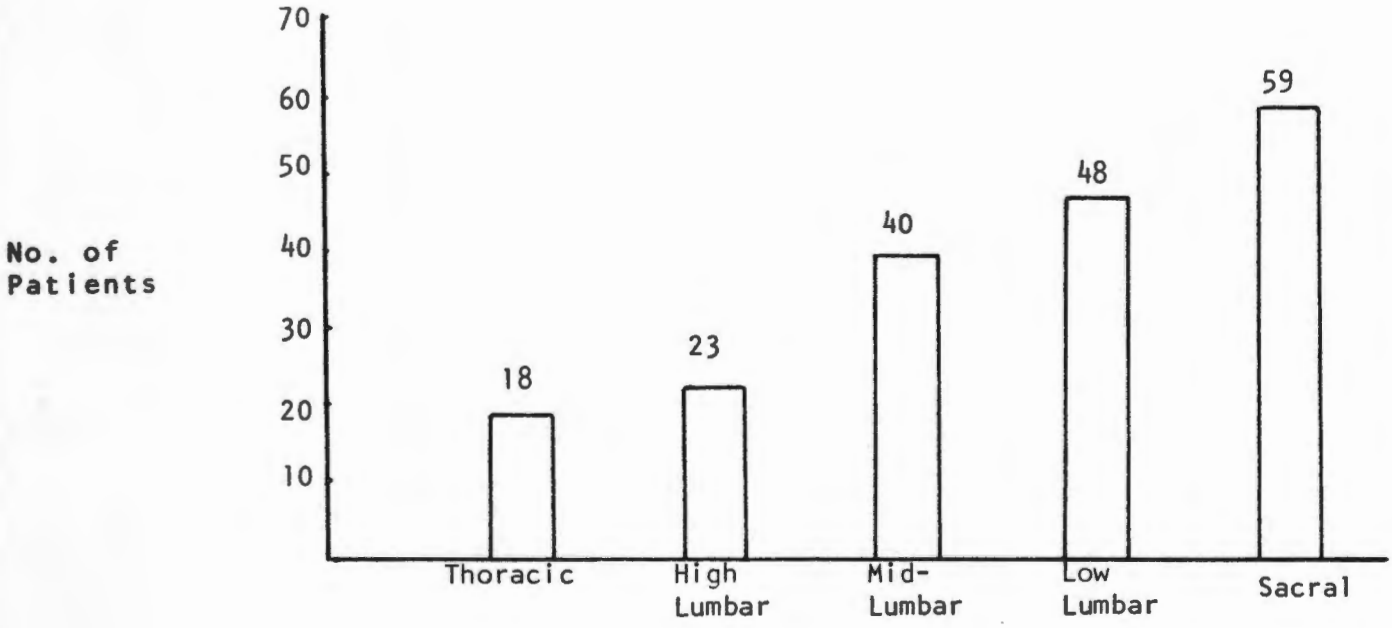
The figure in brackets in the male and female columns is the percentage for each sex in that race group. The figure in brackets behind the total for each group is the incidence of the three race groups expressed as a percentage.

Neurological/...

Neurological Level

The following bar diagram indicates the incidence of the various levels in our series.

FIGURE 20



In asymmetrical lesions the lowest functioning level was recorded and the patient included in that particular group. Asymmetry is taken to be present when there is a discrepancy between the two sides of greater than 2 segments. There were 4 in this series.

FUNCTION

Table 2 indicates the function of all the patients related to level.

TABLE 2

LEVEL	TOTAL	C.W.	F.W.	N.W.	TOO YOUNG
Thoracic	18	1	1	11	5
High Lumbar	23	1	3	12	7
Mid Lumbar	40	17	3	12	8
Low Lumbar	48	33	4	1	10
Sacral	59	50	0	0	9

C.W. - Community walker

F.W. - Functional walker

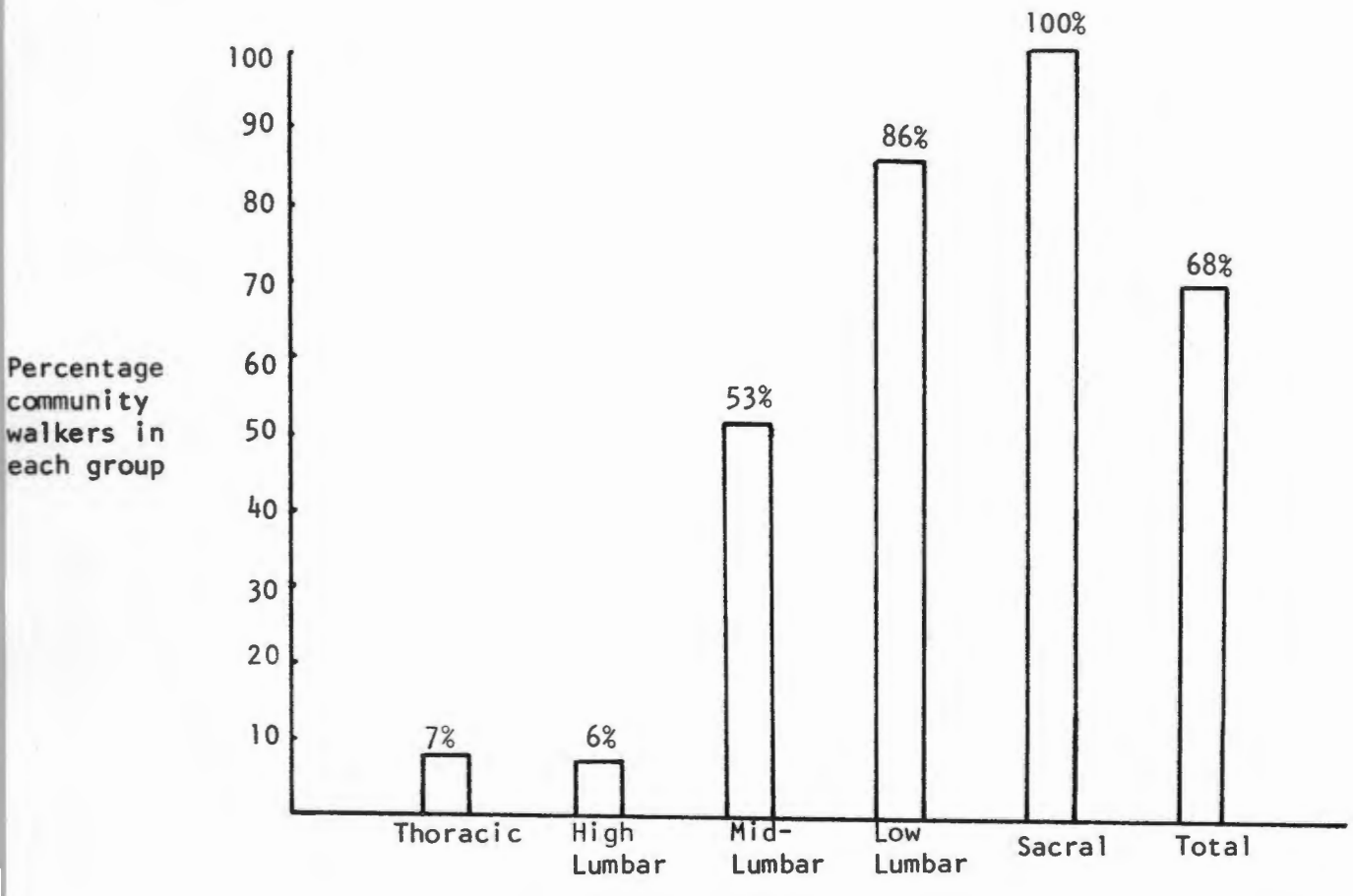
N.W. - Non-walker

Too young : Too young to walk

The/...

The following bar chart (Fig. 21) indicates the percentage of each level that are community walkers. Note, patients that are too young have been subtracted before the calculation and non-functional walkers are included in the non-walker group.

FIG. 21



DEFORMITY/...

DEFORMITY AND TREATMENT

The Spine

Table 3 shows the incidence of the three spinal deformities. The figure in brackets is the percentage of cases with the deformity at each level.

Scoliosis

Only curves greater than 10 degrees have been considered. Of the total of 21 (11% incidence), 16 cases had curves between 10 and 30 degrees. Of the 5 greater than 30 degrees (incidence 2,6%), 1 has had a fusion, 2 wear TLSO braces and 2 are being watched - the curves being just over 30 degrees.

Kyphosis

Cases with 30 degrees and over have been considered. None have had surgery to date and none wear braces.

Lordosis

The primary deformity is corrected if necessary. Lordosis may be present in conjunction with scoliosis i.e. lordoscoliosis in which case anterior fusion and Dwyer instrumentation is the correct procedure - our case was an example of this.

TABLE 3/...

TABLE 3

	THORACIC	HIGH LUMBAR	MID LUMBAR	LOW LUMBAR	SACRAL	TOTAL
Kyphosis	6 (33%)	2 (8%)	1 (2%)	0	0	9 4%
Scoliosis	3 (16%)	8 (34%)	3 (7%)	3 (6%)	4 (6%)	21 11%
Lordosis	0	2 (8%)	1 (2%)	1 (2%)	0	4 2%

The incidence of spinal deformity at each level with the percentage in brackets.

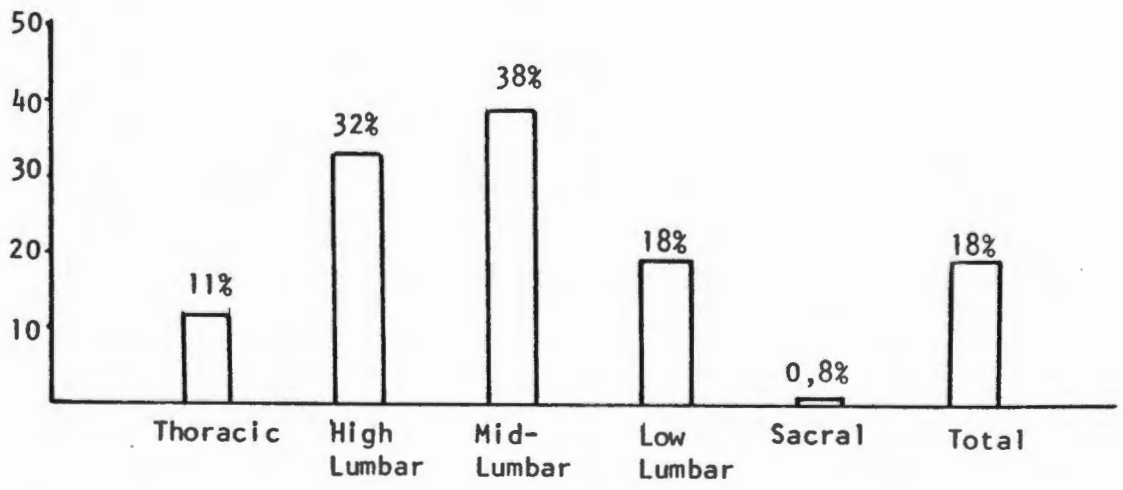
The Hip

Hip dislocation and subluxation are represented in the following table (Table 4) and bar graph: (Fig.22) which indicates the percentage dislocated and subluxed at each level.

TABLE 4

	NUMBER OF HIPS	NUMBER OF PATIENTS
Dislocation	42	27
Subluxation	25	18
TOTAL	67	45

FIG. 22



18% of the total number of hips in the series were subluxed and dislocated.

The/...

The following Table 5 indicates the ambulation status of those with dislocated and subluxed hips:

	DISLOCATED	SUBLUXED
Community Walker	21%	40%
Functional Walker	16%	18%
Non-Walker	63%	42%

The majority of patients with dislocated hips were non-walkers, whereas the majority of those with subluxed hips were walkers.

Deformity/...

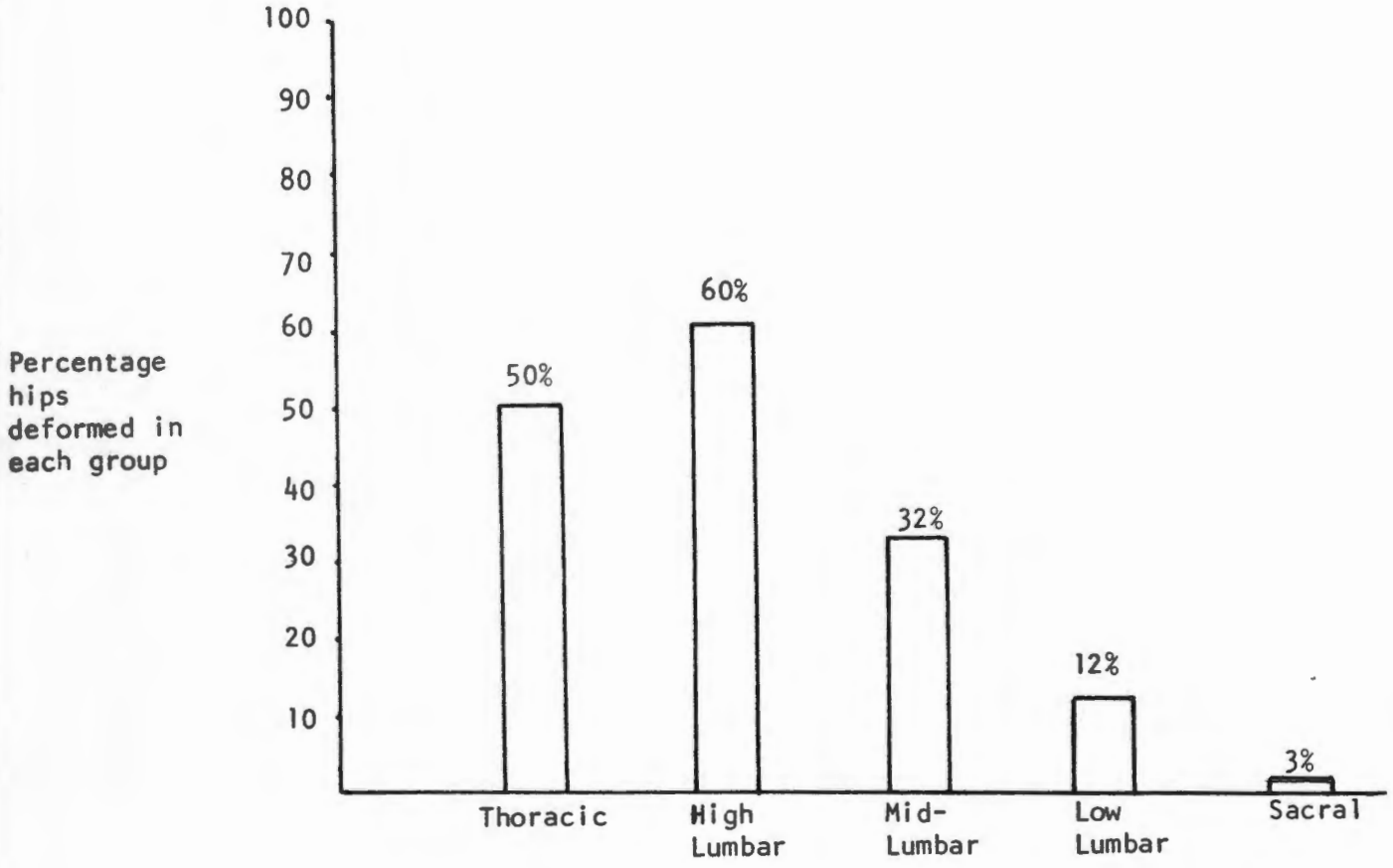
Deformity at the hip without dislocation occurred in the group as follows: (Table 6)

	THORACIC	HIGH LUMBAR	MID LUMBAR	LOW LUMBAR	SACRAL	TOTAL
Number of Deformity	18	24	26	12	4	84
Bilateral	9	10	13	6	2	40
Number of Patients	9	14	13	6	2	44
Percentage of Each Level	50%	60%	32%	12%	3%	46%

Note that this includes all deformities added together. This confirms a higher incidence in the patients with higher levels. The bar chart (Fig. 23) is a graphic representation of the percentage of hips deformed in each group.

Fig. 23/...

FIG. 23



All the deformities in the thoracic group were of the flexion-abduction-external rotation type. At the lower levels they were flexion and flexion-adduction deformities.

Table 7/...

Table 7 illustrates the number of each surgical procedure performed on the hip:

TABLE 7

	NUMBER OF OPERATIONS	NUMBER OF PATIENTS
Open Reduction	4	3
Release of Adductor and iliopsoas	14	8
Release of Flexors	18	9
Release of Adduction Deformity	8	5
Release of Abduction Deformity	1	1
External Rotation Release	1	1
Innominate Osteotomy	3	2
Iliopsoas Transfer (Sharrard)	6	4
Varus Derotation Osteotomy (Proximal Femur)	12	8
TOTAL	67	41

Release/...

Release of adductors and iliopsoas is performed to arrest the progress of subluxation. So far 14 of these procedures have been done. When done early enough subluxation is arrested but still persisted in 10 hips, the remaining 4 eventually reduced. Recurrence of deformity has not occurred in any of them. The oldest patient who has had this procedure is now 4 years of age, the rest are younger. Two were born this year (1985). Taking this into consideration, none of them are community walkers, 2 are functional (household) walkers, 2 are non-walkers and 3 are too young to walk.

Sufficient follow-up has been available for analysis clinically on the following:-
iliopsoas and adductor release, Sharrard's procedure and varus derotation osteotomy. The results are presented in Table 8.

TABLE 8/...

TABLE 8

TYPE OF OPERATION	COMMUNITY WALKER (Excellent)	FUNCTIONAL WALKER (Good)	NON-WALKER (Poor)	TOO YOUNG
Soft Tissue Releases (Includes ilio-psoas and Adductor Recession)	0	5	7	10
Sharrard's Iliopsoas Transfer	2	2	-	-
Varus Derotation Osteotomy	5	1	2	-

Detailed analysis of radiology of the hip has not been performed in this study.

The Knee/...

The Knee

Table 9 indicates the incidence of flexion deformity and recurvatum at the knee:

TABLE 9

	THORACIC	HIGH LUMBAR	MID LUMBAR	LOW LUMBAR	SACRAL	TOTAL	NUMBER PATIENTS
Flexion Deformity	16 (44%)	8 (17%)	10 (12%)	14 (29%)	2 (3%)	50	25
Recurvatum	0	4	6	0	0	10	5

Many of these deformities occurred in the first year of life and thus were not surgically corrected as spontaneous improvement occurred or a mild residual deformity persisted of less than 20 degrees which allows ambulation.

Hamstring release was performed on 7 cases where ambulation in calipers was being hampered.

Wheelchair users i.e. the thoracic and high lumbar cases with the deformity were not treated.

Recurvatum deformity has not been treated surgically by us as the extended position is suitable for the fitting of calipers and ambulation.

The Foot/...

The Foot

The incidence of the various deformities are as follows:

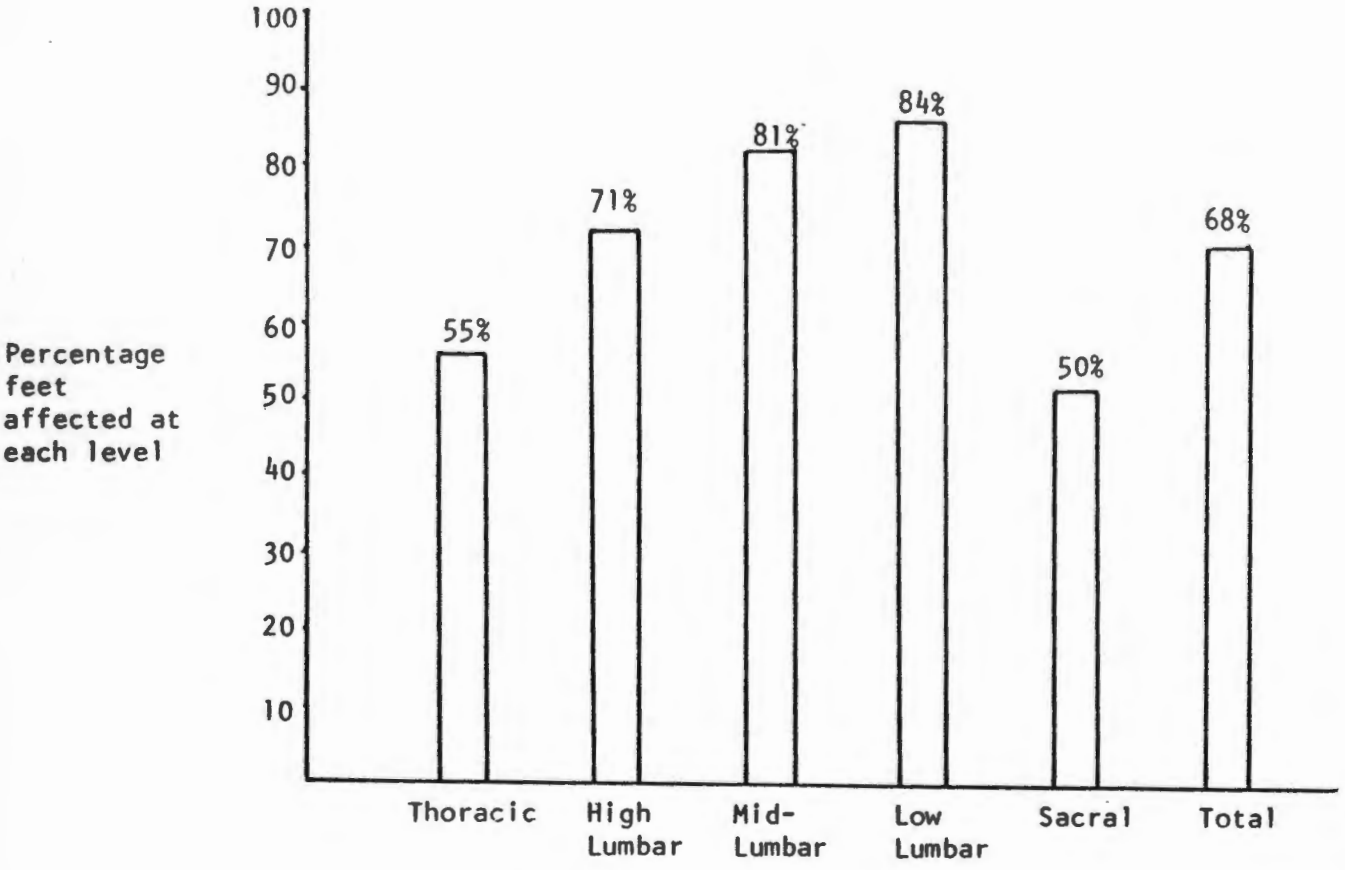
TABLE 10

	NUMBER OF DEFORMITIES	BILATERAL	NUMBER OF CASES
Calcaneus	105	45	60
Valgus	36	15	21
Vertical Talus	7	2	5
Equinovarus	60	26	34
Equinus	23	8	15
Varus	5	0	5
Metatarsus Adductus	3	1	2
Cavovarus	12	5	7
Claw Toes	6	3	3
TOTAL	251		

40 Patients out of the study group were without foot deformity (68% of feet were deformed). 148 Patients had 257 deformities between them, 105 being bilateral. 17 Patients had asymmetrical lesions, for example calcaneus one side, and equinovarus the other. 13 Patients had unilateral deformity.

Fig. 24/...

FIG. 24



Foot deformity is common at all levels affecting over 50% of each group and 68% of the total.

This bar diagram (Fig. 24) indicates the incidence of foot deformities at each level illustrated as a percentage of the total number of feet in the series i.e. of 188 x 2 = 376 feet.

TABLE 11/...

TABLE 11

	NUMBER OF OPERATIONS	NUMBER OF CASES	REPEAT
Posterio-medial Release	37	23	11
Tendo-Achilles Tenotomy	37	21	
Posterior Release	2	2	
Dwyer Valgus Osteotomy	5	3	
Steindler Plantar Fascia Strip	4	3	
Triple Arthrodesis	4	2	
Tendon Transfer (Miscellaneous)	13	10	
Tibialis Anterior Transfer (Peabody)	36	20	
Correction of Vertical Talus	7	4	
Extensor Release	8	4	
Grice	10	6	
Curettage Pressure Sores	15	8	
Jones' Correction of Clawed Big Toe	3	2	

This table indicates the frequency of the various foot operations performed. Postero-medial releases had to be repeated on 11 feet after the deformities recurred i.e. 29%. Peabody's transfer was found to be a very useful procedure as all but 3 cases remained plantigrade. These 3 developed equinus deformities and had to have elongation of the transferred tendon!

TIBIAL/...

TIBIAL TORSION

Only 1 case with a thoracic lesion had external tibial torsion bilaterally.

Internal tibial torsion occurred bilaterally in 12 patients and unilaterally in 1. They were all mid-lumbar or low lumbar lesions. Assessment of this condition is carried out clinically only.

Derotation osteotomy was performed on 12 tibias with internal tibial torsion on 7 patients.

Mortality

One patient aged 16 months died during the study period. Four other deaths occurred prior to the start of the study, all being under 6 months of age.

Fractures

Only 4 fractures that we know of were recorded during the course of the study.

DISCUSSION/...

DISCUSSION

Survival of these patients certainly has improved thanks to the advance of technology and expertise. As far back as 1964 Sharrard reported that the mortality in the first year of life had improved from 90% to 40%. These figures are very much smaller today. Professor Peacock reports on 4% deaths in a group of 130 patients he has treated at the Red Cross War Memorial Children's Hospital; all of these died in the first month of life. (Personal communication). Only one of the number of cases we have been studying since 1983 has died.

Orthopaedic management aims at the correction of deformities to allow ambulation when practical or else to facilitate wheelchair sitting and hygiene. We have adopted the attitude which is developing elsewhere (Menelaus 1980) (Schaffer 1983) which is to do simple muscle releases to correct deformity. We share the view that major surgery is frequently non-productive.

Sex and Racial Factors

Cape Coloured people comprised the largest race group seen at the clinic. This is not in accord with the expected small incidence indicated by the Cornell (1983) study.

The incidence of the disease is higher in males in whites and coloureds but reversed in blacks. In Menelaus' series (Caucasians) it was commoner in females. These findings are

only/...

only of significance in definitely precluding a sex-linked genetic factor.

Last Functioning Level

The lower lesions are far commoner than higher lesions, a factor favouring ambulation. This is the experience of others too. (Menelaus 1980, Sharrard 1979).

Function

68% of the group old enough to walk are community walkers, which is to be expected considering the higher incidence of lower lesions.

The Spine

Kyphosis

The overall incidence of kyphosis is 4% (33% in thoracic lesions). These figures are in accord with others' findings. Sharrard (1979) gives an incidence of 5% overall and 10% in thoracic lesions. Banta (1976) had a 10% incidence overall.

None of our kyphotics have had surgery because none have recurrent ulceration.

Scoliosis

The incidence of scoliosis in our series is lower than in other reported series. Banta (1976) in his series had an incidence of 50% at 15 years of age compared to

none/...

none in our series. Only 6 patients have reached this age and thus comparison is not realistic.

The incidence of scoliosis of greater than 30 degrees in the group as a whole is 2,6%. This figure is less than the one quoted by Menelaus (1980) for sacral lesions at one year.

The incidence will probably increase in this series as greater numbers reach maturity. This study should be repeated on the same patients after 10 years to throw light on this point.

Only one patient has had scoliosis which increased in severity in spite of bracing, thus requiring surgery.

Hip

The incidence of hip deformity and dislocation is 33% in our series. Hip subluxation and dislocation per se occurred in 18% of patients. In other series (Sharrard 1964) (Schaffer 1983) it varies from 30-50%.

Dislocation and subluxation tended to occur in the high lumbar (63%) and mid-lumbar (38%) groups. Sharrard (1979) had the highest incidence in L1-L4 groups which correlates with the findings of this study. This supports the hypothesis that flexion and adduction deformity would occur at this level and predispose

to/...

to subluxation and dislocation.

Hip deformities per se were commonest in thoracic, high lumbar and mid-lumbar lesions tailing off in the lower levels.

We are conservative in the management of dislocated hips in SBC, only 4 open reductions were performed.

37% of the group with dislocated hips and 58% of those with subluxed hips were able to walk.

Of 84 patients with hip deformity, 42 had soft tissue releases. Many of the deformities were of insufficient severity to warrant surgery, others were in non-walkers.

The efficacy of iliopsoas and adductor release has not been fully assessed in these patients: this analysis should take place as a separate study. The results I have presented are based on a short-term clinical impression only. Schaffer (1983), reviewing the literature, reports varying stability of the hip from 20 - 90% following this procedure.

Knee

The incidence of knee deformities over 10 degrees is 16% in this series, which is much lower than Menelaus' (1980), 70%.

Hamstring/...

Hamstring release only, performed at the time of ambulation, has proved a satisfactory way of dealing with a flexion deformity. Serial plasters have no place in the treatment because of the problems with anaesthetic skin. Sharrard (1979) prefers to treat the knee before the foot but stands alone as Schaffer (1983) and Menelaus (1980) have the same approach as we do i.e. to treat the foot soon after birth. The knee deformity should be left until after the first year if possible as many will resolve spontaneously.

Foot

Foot deformities occurred in 68% of feet in our series. Menelaus (1980) gives a figure of 94% and Schaffer 90%. Sharrard (1968) 82%. It is not possible to explain why our figure is lower than that of the others.

Calcaneus deformities are the commonest in our series. Schaffer (1983), Sharrard (1979) and Menelaus (1980) had equinovarus as the commonest deformity.

We have found that proper timing of surgery is essential. It should be carried out when standing has started to ensure maintenance of the correction. This is perhaps why Janda (1984) had such poor results with Peabody's transfer - 100% failure in 6 cases. The procedure gave a plantigrade foot in all but 3 of the 37 Peabody procedures done in our unit. In these, overaction

of/...

of the transferred muscle caused equinus deformity. Menelaus (1980) has had similar results if tibialis anterior alone (Peabody's transfer) is transferred. He has found that adding other tendons produces valgus or varus deformities. Sharrard (1968) transfers peroneus brevis in addition and has had good results.

Equinovarus remains a challenge with a 29% recurrence rate in our series, 30% in Menelaus' (1980), and 20% in Sharrard's (1968). The deformity tends to be very rigid due to spasticity of the deforming muscles - Stark and Baker type 11 lesions. Our approach is to do nothing to these feet until ambulation starts. Circumstance has largely dictated this approach as early corrective plaster changes would improve the deformity, diminishing the magnitude of eventual corrective surgery. An effort should be made to obtain facilities to use Plaster of Paris in the clinic and to treat those that live nearby with serial plasters.

Calcaneus deformities are less rigid (Schaffer 1983) indicating their occurrence in Stark & Baker type 1 lesions. This makes correction much simpler and reduces the recurrence rate.

CONCLUSION

This study has shown that our experience in Cape Town is very similar to that of other authors abroad. The size of this study group is much the same as that of others reporting their experience. (Menelaus 1980, Schaffer 1983, Sharrard 1979). The reason for the small size of their reports is to allow for long-term follow-up. This feature is lacking in our study and I think the smaller incidence we have of many deformities is because of short-term follow-up. The average age of the study group is 6,5 years. It would be ideal to study this group when they have all reached maturity. Then only will true comparative results be available. Nevertheless, what we have is a good guide to what is happening to our patients with the present approach to management.

It is recommended that the following in-depth studies should be performed in the near future:

(a) our experience with the management of foot deformities, our success with Peabody's transfer for calcaneus and our management of equinovarus deformity

(b)/...

(b) our approach to flexion-adduction contracture of the hip by using the iliopsoas and adductor tenotomy - a prospective study comparing this with iliopsoas postero-lateral transfer would be interesting, though I think it would be morally indefensible to subject patients to this major procedure

(c) review of the incidence of scoliosis in 10 years time.

It is important to draw up a proforma for each of these studies starting now. This will provide a more accurate comparative study of the various aspects in a few years time.

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