

IMAGING OF TUMOURS OF THE URINARY TRACT IN CHILDREN,
WITH PARTICULAR REFERENCE TO WILMS' TUMOUR

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DEDICATION

To fellow travellers who have tried to
look behind the image.

"WE SHALL NOT CEASE FROM EXPLORATION
BUT THE END OF ALL OUR EXPLORING
WILL BE TO ARRIVE WHERE WE STARTED
AND KNOW THE PLACE FOR THE FIRST TIME."

T.S. Eliot. "Little Gidding"

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STATEMENT OF CANDIDATE

I declare that the work on which this thesis is based is original (except where acknowledgements indicate otherwise), and that neither the whole work nor any part of it has been, is being, or is to be submitted for another degree in this or any other University.

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ABSTRACT

Abstract

The investigation of an abdominal mass in a child is a common problem in the radiology department of the Red Cross Children's Hospital. The majority of these masses involve the urinary tract. The commonest neoplasm is a Wilms' tumour of the kidney.

Against a pathological and clinical background the investigation of Wilms' tumour by diagnostic imaging is presented. The imaging modalities currently utilised are the intravenous urogram (IVU), ultrasound (US), computed tomography (CT) and magnetic resonance (MR).

Using the material available in the last decade, the principles, techniques and imaging characteristics of these modalities are investigated and compared. These results are reflected against those reported in the medical literature. This literature is not yet extensive as the current technology has only been available for the last six to seven years.

The IVU has in the past been the main imaging modality and we still use it extensively. Its strengths and weaknesses are discussed. In the last five years US has taken its place as the primary method of diagnostic imaging. We have found that with our increasing experience that this is justified. The use of US and IVU in a practiced hand is a powerful diagnostic combination. CT as a primary investigation is not readily available at our institution. We have used it for comparative purposes in about 20% of our recent

cases. CT has not added greatly to our initial diagnostic impression. However, it has been most useful for follow up of metastasis and for assessing the normality of the lungs before ceasing chemotherapy. Our experience with MRI is limited and confined to unusual presentations in the last year. Other modalities such as arteriography and nuclear medicine have special indications which are to be discussed. The remaining tumours of the upper urinary tracts are all rare, but are reported and the literature researched.

In the lower urinary tract the main pelvic lesion is a rhabdomyosarcoma. The comparative advantages of the IVU, US, CT and MRI are also noted.

In the pelvis, US has also become the primary imaging modality, and is replacing contrast medium cystography. However, examples of the latter are included as it still has a place, particularly in the less sophisticated institutes. CT and MRI, when available, have imaging advantages in the pelvis and are becoming the methods of choice for follow up.

The main objective of this document has been to investigate the available imaging techniques, but, against this overall theme, the clinical care of the child is most important. With this in mind the treatment protocols that are used at our hospital are noted in the appendices to the thesis.

INTRODUCTION

1. The general content

In its broadest sense diagnostic radiology has now developed into the science of imaging. Conventional radiology that employs ionising radiation has been refined by computed technology to produce images of increasing depth and definition. Ultrasound and magnetic resonance using non ionising forms of radiation to create images have come to the forefront as tools of the radiologist.

The aim of this thesis is to ascertain the values of these imaging modalities with reference to tumours of the renal tract in children.

The objective is to evaluate the various tumours and problems that accompany them. In the majority of cases we have approached this from a clinicopathological aspect. We have examined the current imaging modalities and the available literature and noted the effectiveness of these methods, both in our hands and those of others. It must be recorded that the technical advances that have made real time ultrasonography and fast speed computed tomography a boon to paediatric practice have taken place fairly recently (since 1979). The literature on the subject has not yet had time to accumulate and exhaustive comparison of modern imaging is not yet possible. Nevertheless we have been able to form an opinion about them.

The material is my own personal experience in paediatric radiology. This extends over 25 years but

has been confined to the imaging modalities in use at the Red Cross Children's Hospital during the last decade. The material mostly concerns Wilms' tumour. There have been 69 cases at our hospital in the eleven and a half year period from 1975 to June 1986. It was only at the start of the eighties that the technical advances in ultrasound and computed tomography made their impact on paediatric imaging. We have therefore confined the figures analysed for ultrasound and computed tomography to the period 1981 - 1985. During this period there were 27 cases.

The method has been to review the available imaging modalities and to select appropriate illustrations. If there is to be merit in this work its strength must lie in the quality of these illustrations. They should speak for themselves and we have tried to label the relevant points. The words of an old adage state that a good picture is worth a thousand words. They have enabled me to shorten the text and I hope make it simpler to read.

My interest in writing about this subject arose when I collected material for the relevant chapter in the textbook "Practical Paediatric Uroradiology" published in 1985. This was written in conjunction with Ian Aaronson, our paediatric urological surgeon. For this thesis I have started afresh, approached the problem from a much broader perspective and with few

exceptions, collected an entirely new set of cases. An initial problem in the presentation was to put relevant illustrations into the compartments that applied to their modality. I decided against this. Therefore computed tomographic images are scattered out of their section. For the most part, the modalities that were primarily used or best showed the lesion have been grouped around a case as illustrations. The virtue of this method is to show the comparison they would have in clinical practice. In the majority of cases the illustrations follow the page in which they are referred to and concern the relevant aspect of the tumour. We have covered most aspects with our own material. Where we have had to make an exception and borrow a case, such as in the condition of nephroblastomatosis we have inserted an out-of-sequence computed tomogram in the ultrasound section.

I have also tried to give a short resume of the principles behind the main methods of imaging. To go into detail would be beyond our objectives. However, these sections might be considered either too short or too long. That on magnetic resonance is in fact a compromise to confine it to the urinary tract.

2. The general concept

The problem of urinary tract tumours in children is relatively common and the standard textbooks on Paediatric Surgery (Swenson; Mustard et al; Holder & Ashcroft) were consulted. Most of the abdominal masses

of surgical concern arise from the retroperitoneal structures and roughly two thirds of them are of renal origin and concern either hydronephrosis or Wilms' tumour. The other common tumour is neuroblastoma.

Figures from the Los Angeles Children's Hospital (Mustard et al) show that these three entities covered 75% of retroperitoneal masses. The incidence figures were hydronephrosis 36%, Wilms' tumour 20% and Neuroblastoma 19%.

The differentiation of hydronephrosis from Wilms' tumour should not be difficult. Firstly, some 90% of our hydronephrosic masses present in the first few months of life. This is an age when a Wilms' tumour is uncommon. Secondly, ultrasound in either prenatal or postnatal periods clearly differentiates the cystic entities from solid tumours.

The differentiation of Wilms' tumour from neuroblastoma is not always so simple. In the abdomen the majority arise in close relationship to the kidney. As a source of diagnostic error this will be discussed, but neuroblastoma is not a feature of this thesis. Suffice it to say that any experienced radiologist must know that the differentiation of neuroblastoma from Wilms' tumour should be a clinical one in the first instance. Children with neuroblastoma are usually sick, fretful and feverish compared to the Wilms' tumour child who commonly presents with an asymptomatic abdominal

mass.

From these general statements it emerges that Wilms' tumour is by far the most common and important of the urinary tract tumours in children. It will occupy just over two thirds of the bulk of this thesis. The problem will be approached by first presenting the pathological and clinical aspects of the tumour. Against this background the imaging modalities and techniques will be described.

For the remainder of the thesis, imaging of the other tumours of the urinary tract will be evaluated with the caveat that it is unlikely that any radiologist will see them all during a working lifetime. We have therefore not burdened the reader with overelaboration in this section. It becomes obvious that a radiologist should use the modality with which he is most familiar, but that he must keep abreast of the times.

Finally, although treatment is not the main concern of this document, its effectiveness must be the ultimate goal of all early diagnosis. Therefore, the staging and prognosis of this interesting tumour is outlined and treatment protocols given in the appendices.

UPPER TRACT TUMOURS

=====

Wilms' Tumour (Nephroblastoma)

Wilms' tumour is not only the commonest renal malignancy in children but is the commonest eponym in renal oncology. It is named after the German surgeon Max Wilms (Fig.1) who was an international figure in nephrology at the turn of the century. He held the Chair of Surgery at the University of Heidelberg and died in 1918 of diphtheria at the age of 51 years after operating on a French prisoner of war. Paradoxically the patient recovered! (Beighton & Beighton). Although there were earlier descriptions notably by another German, Rance in 1814 and the Edinburgh physician Ebenezer Gairdner in 1828 (quoted by Mustard et al) the treatise by Wilms on four cases in 1899 clarified the pathology. This was the first account that classified the neoplasm as an embryonal sarcoma. It is only in recent years that this classical concept has been modified by the collective approach from the National Wilms' Tumour studies.

Wilms' tumour has a fairly uniform incidence in children throughout the world and about 450 cases a year are reported from U.S.A. It constitutes 6.2% of all cancers in white American children and 7.9% of cancers in black children (Young & Miller). Its main incidence is between one and five years with a peak at three years (Belasco et al).

The vast majority of cases are sporadic, but



MAX WILMS 1867 - 1918

Max Wilms, the son of a lawyer, was born near Aachen in 1867. He studied at a number of German universities, obtaining his doctorate at Bonn in 1890. Wilms was then appointed to the Pathological Institute of Cologne, where he spent several years before moving to Leipzig for training in surgery under Professor Trendelenburg. In 1907 he was appointed professor of surgery in Basle and in 1910 he reached the peak of his career when he was called to the chair at the University of Heidelberg.

In 1899 while working at the Institute of Pathology, Wilms produced a monograph on the pathology of mixed tissue tumours. Although he was only 32 years of age he had been interested in histology of neoplasms of this type for nearly a decade. His book dispelled existing confusion and his classification provided the foundation for modern concepts concerning this group of disorders.

Acknowledgements Prof. P. Beighton.

perhaps one to two percent have a familial incidence. It is important to recognise these familial cases, not only for genetic counselling, but for early diagnosis in suspected families (Knudson & Strong).

The condition is uncommon though well documented in adults (Babaian et al). It is debatable whether these adult cases represent true nephroblastomas or sarcomatoid renal carcinomas.

Pathology

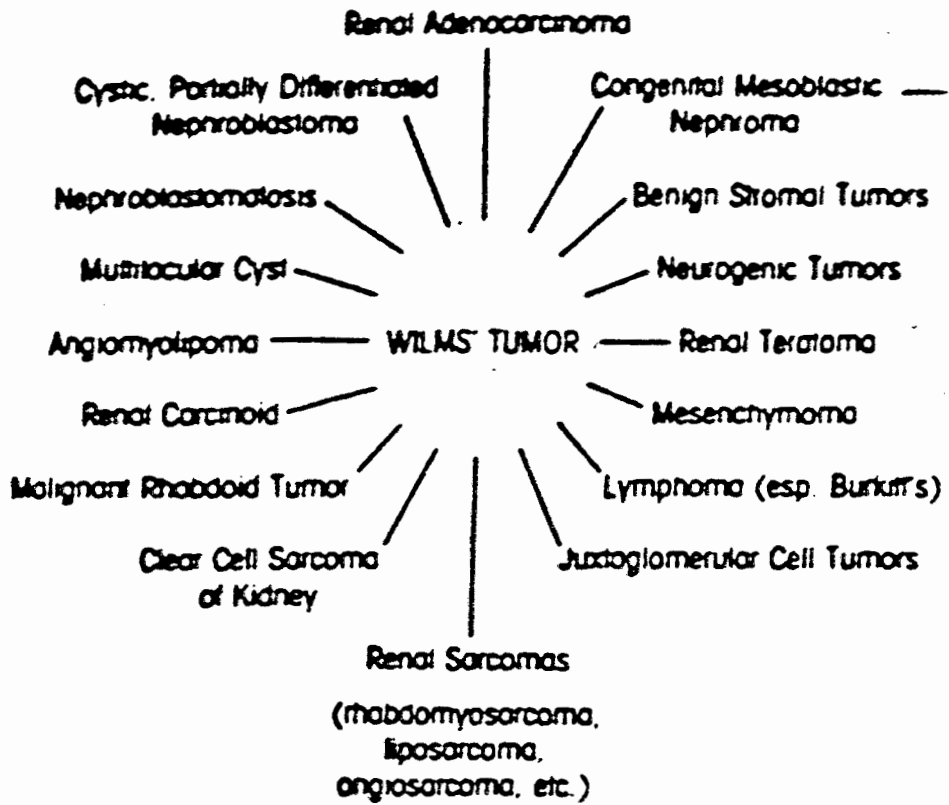
The classic Wilms' tumour is a solitary lesion that occurs in any part of the kidney. It is usually solid and fleshy with a pseudo capsule that separates it from the rest of the kidney. Areas of haemorrhage and jelly-like necrosis are frequent so that it may develop a cystic appearance. The tumour frequently remains silent and can grow to a massive size replacing much of the kidney tissue and penetrating into the renal pelvis. This may cause "non-function" or more correctly non-visualisation on the intravenous urogram. Venous invasion is also a frequent feature of the tumour and both infiltration of peripelvic and surrounding soft tissue with lymph node involvement may occur.

In order to get a collaborative multicentre approach to therapy the first National Wilms Tumour Study (N.W.T.S.-1) was launched in 1969 and has been updated by two further studies. These have been published in 1981 and 1984 by D'Angio et al. These studies have done much to clarify our knowledge about

renal neoplasms and to provide a uniform and successful approach to their treatment.

Nephroblastoma may be described as an embryonic neoplasm, but modern pathology has shown it to be an extremely complex tumour. It is a triphasic tumour with epithelial, blastemal and stromal elements. Usually the tumour is mixed in nature with differentiated epithelial tissue forming glandular acini but sometimes there may be a large predominance of either blastemal or fibromyoid stromal elements (Chatten). The non-epithelial elements may further differentiate into striated muscle, adipose tissue, cartilage or bone. The pathological dilemma increases when one considers that most primary renal tumours must be of blastemal origin and are therefore all related to Wilms' tumour. The reader is referred to Table 1 from Beckwith in which the spectrum of intrarenal neoplasms that are related to Wilms' tumour is illustrated.

From clinicopathological correlations at least three "unfavourable" tumour types emerge (Beckwith & Palmer). These comprise only 10% of cases but are responsible for over 60% of deaths. There is an anaplastic type that tends to occur in older children and a rhabdoid type that tends to occur more often in infants (Belasco et al). However it is the third "unfavourable" type that is of particular interest to radiologists. This is the clear cell sarcomatous type



Reference: Beckwith JB

which is liable to have bone metastases (Marsden et al; Morgan & Kidd). These are more or less monophasic tumours in which the descriptive term of rhabdomyosarcomatoid with clear celled and hyalinizing patterns is used (Lanzkowsky). The subject is still under debate and it has recently been suggested that the rhabdoid and clear cell types be removed from the Wilms' tumour spectrum (Kaufman). Recognition of these unfavourable types is not only important to radiologists but to the oncologist as well. Staging of tumours influences the decisions about the use of radiotherapy but unfavourable types influences the addition of drugs such as adriamycin in the treatment.

Related to the more favourable types are the three entities of multilocular cyst, congenital mesoblastic nephroma and nephroblastomatosis. The multilocular cyst is an uncommon renal neoplasm that consists of a localised, well circumscribed and encapsulated renal mass. It is characterised by multiple cysts that are usually non communicating and lined by epithelial cells with a distinct stroma (Beckworth & Kiviat; Madewell et al). There may be different forms of this entity as it has a biphasic age and sex distribution affecting boys in childhood and women in adulthood (Madewell et al). Although it is usually classified as a benign neoplasm recent reports show a definite incidence of Wilms' tumour in the lesion. The current trend is to classify the lesion as a multilocular cystic nephroma and to

place it at the benign end of the Wilms' tumour spectrum.

Congenital mesoblastic nephroma is a firm solitary renal mass that manifests itself early in life. It grossly and microscopically resembles a leiomyoma or low grade leiomyosarcoma that contains trapped nephrons (Bolande). There is still considerable debate as to whether this is a separate benign hamartoma or a monomorphous Wilms' tumour variant.

Nephroblastomatosis

Nephroblastomatosis is thought to occur from an arrest of normal nephrogenesis so that there is a persistence of primitive metanephric blastema. The normal sequence of development is that the kidney develops from two precursors. The mesonephric (ureteric) bud forms the ureter, renal pelvis and calyces. The metanephric blastema forms the renal parenchyma that contains both stromal and epithelial elements. This differentiation has normally taken place by 34 - 36 weeks gestational age. These areas of nephroblastomatosis may occur singly or together. Much of our knowledge of this condition comes from the work of Bove and McAdams. The condition may be interpreted as a precursor of Wilms' tumour, but it may be present with or without development into Wilms' tumour, either in the same or opposite kidney. It is however considered that when nephroblastomatosis is found in the

contralateral kidney of an affected child there is a greater likelihood of these developing into a Wilms' tumour. Persistence of this embryonic tissue not only occurs with Wilms' tumour (23 examples of multifocal nephroblastomatosis in 69 patients with nephroblastoma) but also in patients with hemihypertrophy, Beckwith-Wiedemann Syndrome and major chromosomal abnormalities. (Bove & McAdams).

The condition has been divided into diffuse and multifocal forms. The diffuse is less common and less likely type to be associated with Wilms' tumours. The diffuse form has two sub-divisions. a) A paracortical (infantile) type in which the kidneys are greatly enlarged and the entire renal parenchyma consists of metanephric blastema. There is no normal tissue and in its true form the condition is usually incompatible with life (Franken et al). b) A superficial diffuse (late infantile) type in which the persistence of metanephric blastema is subcapsular. The radiographic appearance of this is either symmetrical or asymmetrical enlargement of the kidneys (Tealander et al).

The multifocal form is the more common and more commonly associated with Wilms' tumour. There are various sub-types but nodular renal blastema, sclerosing metanephric hamartoma and Wilms' tumourlets are considered the most significant. They are considered non-invasive but potentially malignant (Machin). The Wilms' tumourlets are monophasic in nature and contain

only epithelial cells. The nodules may be up to 3cms in size and as such they should be theoretically detectable by imaging.

We have not recognised this condition by our imaging methods. A neonatal case has been reported by Rosenfield et al in which both kidneys showed large avascular areas on arteriography. A more recent report is by Montgomery et al who reported that these areas could be recognised either as relatively anechoic on ultrasound or nodules of diminished attenuation on computed tomography. As we have no proven experience of the condition, we report these facts at this stage and note that the only images we have are by courtesy of Montgomery et al and are shown in Fig. 14.

Staging

To appreciate the benefits of early diagnosis the radiologist must have a working knowledge of the grading of Wilms' tumour. One of the results of the National Wilms' Tumour Studies (N.W.T.S.) was the opportunity to simplify, unify and evaluate methods of grading and treatment. The grading of the N.W.T.S. has been widely adopted and was updated by D'Angio in 1981 and as shown in Table II. This modification includes nodal involvement and tumour spillage as Stage II involvement.

Clinical features

Most children present with a painless abdominal mass often discovered accidentally by the mother when

TABLE II

STAGING SYSTEM FOR NATIONAL WILMS'
TUMOR STUDY (N.W.T.S.2)

Stage I

Tumour limited to kidney, completely excised. Capsular surface intact; no tumor rupture; no residual tumor apparent beyond margins of excision.

Stage II

Tumor extends beyond the kidney but is completely excised. Regional extension of tumor; vessel infiltration; tumour biopsied or local spillage of tumor confined to the flank. No residual tumor apparent at or beyond margins of excision.

Stage III

Residual nonhematogenous tumor confined to the abdomen. Lymph node involvement of hilus, periaortic chains or beyond; diffuse peritoneal contamination by tumor spillage; peritoneal implants of tumor; tumor extends beyond surgical margins either microscopically or macroscopically; tumor not completely removable because of local infiltration into vital structures.

Stage IV

Deposits beyond Stage III (e.g. lung, liver, bone, brain).

Stage V

Bilateral renal involvement at diagnosis.

Reference: D'Angio et al

bathing her child or by the physician on a routine examination. The mass may become large before it is investigated and sometimes may only present after minor incidental trauma has drawn attention to it. The mass is hard rather than firm and when it extends across the midline may be difficult to differentiate from a neuroblastoma. In practice children with Wilms' tumour often appear fit and well, whereas neuroblastoma cases are sick, feverish, fretful and may have other stigmata such as proptosis. Only about 15% have haematuria and other features such as malaise, anorexia, anaemia and hypertension may be vague and non-specific.

The diagnosis should be suspected in all children who present with an abdominal mass, hypertension or haematuria. The usual imaging problem is differentiation from neuroblastoma but other mass lesions such as hydronephrosis, cystic kidney disease and rarer forms of renal neoplasm may have to be considered.

Associated Abnormalities

About 15% of Wilms' tumour are associated with congenital anomalies (Lemerle et al; Pendergrass). These include:-

Aniridia

There is no association with familial autosomal dominant aniridia. In the sporadic form in which there is a severe hypoplasia of the iris there is an increased incidence of Wilms' tumour. The exact range of

incidence is uncertain but varies from 1 in 75 (Miller et al) to 1 in 3 (Pilling). In this syndrome there is a consistent deletion in the short arm of chromosome 11 (Yunis & Ramsey). The fully expressed syndrome includes development of tumour before 3 years of age with congenital eye lesions, mental retardation, genitourinary anomalies and deformities of the pinna (Haeken & Miller).

Hemihypertrophy and Growth Disorders

There is an increased incidence of all embryonal cancers, including nephroblastomas, adrenal cortical neoplasms and hepatoblastomas in children with hemihypertrophy. Wilms' tumour may appear in either the ipsilateral or contralateral kidney and there may be other associated features such as multiple pigmented nevi and haemangiomas (Fraumeni et al). An increased incidence of Wilms' tumour has also been reported with neurofibromatosis (Stay & Vawter). A case in a black child with neurofibromatosis has been reported by Walden et al, but we have not encountered the association. We have had a case in the Beckwith-Weideman syndrome (macroglossia, gigantism and exomphalous) but this is a well-known association (Tank & Kay).

Genitourinary Abnormalities

There is an undoubted overall increased incidence (4.4%) associated with developmental disorders of the renal tract (Pendergrass). These include male

pseudohermaphroditism (Drash et al) gonadal dysgenesis, defects in kidney fusion, ectopia, duplication of the collecting system, hypospadias and cryptorchidism. Although it has been reported in the fairly common developmental anomaly of horseshoe kidney (Shashikumar et al) we have not noted this in our 130 cases and this reflects the experience of other large series (Gay et al).

Examination

Laboratory Studies

Our routine protocol for children with retroperitoneal masses includes the following examinations:

1. Blood: Full blood count. Blood chemistry, base line tests for kidney function that include blood urea nitrogen and creatinine.
2. Urine: Urine analysis and urinary catechol amines when neuroblastoma is suspected.
3. ECG and echocardiogram for all patients who will be treated with adriamycin.
4. Chromosome karyotyping; if relevant when other congenital anomalies exist.

Imaging

The imaging modalities that may be used for the diagnosis and follow up of Wilms' tumour are:-

- 1) Conventional radiology with the intravenous urography (IVU) as a keystone to diagnosis.
- 2) Ultrasound (US)

- 3) Computed Tomography (CT)
- 4) Magnetic Resonance Imaging (MRI)
- 5) Nuclear Medicine (radionuclide scans) (RNS)

Such richness of choice may present the clinician with a dilemma but the whole battery is not required. The sophisticated technology used to acquire data for computed images is expensive. However it is precise and may be more cost effective in such situations as the CT follow up of pulmonary complications. Hospitals having CT on site make use of US for the primary renal examination (Kuhns, Kuhn & Berger) and if this is positive perform abdominal and chest CT at the same time. Many smaller children's centres in the U.S.A. and large ones in less affluent countries have reasonable access to CT but do not have it on site. For the latter radiologists, the simpler modalities of ultrasound and IVU are recommended. In practiced hands a combination of clinical acumen, US and a well performed IVU are highly effective and should detect both the intrarenal and morphological nature of the mass. We perform the US first and when this is positive, follow it by an IVU. This local preference for an IVU acknowledges the fact that hard copy US images are not always easy to interpret by any other than their creator. In addition many surgeons prefer a spatial impression of the tumour on the IVU and the visual confirmation that it is not bilateral. It must follow that less use of the IVU will

be made in the future and that a recent book on paediatric oncology imaging (Sty, Hernandez et al) has omitted any account of it in the text. However the IVU is a relatively simple examination and in many countries is still a major investigation for abdominal masses. It would be a pity to breed a new generation of radiologists who are inexperienced in interpreting its virtues.

In the description of imaging techniques that follows it would be an error of judgement if all the relevant illustrations were confined rigidly to their sections. In order to compromise and attempt to achieve the best of both worlds we have jumped ahead a little and have combined many of the IVU and US illustrations together and included CT examinations when relevant. This will present some inconvenience, but for those whose experience is limited it may be an advantage. They will be able to compare and evaluate the images. This is an objective of the thesis.

THE INTRAVENOUS UROGRAM

Intravenous Urogram (IVU)

For many years the IVU was the main method of diagnosing Wilms' tumour. Its recorded accuracy is in the region of 90% (Mustard et al). The figures from a large study by D'Angio et al and a smaller one by Ehrlich et al were analysed and combined with those from the International Society of Pediatric Oncology by Lemerle et al.

This research on major co-operative studies was recorded by Belasco et al. They showed that the preoperative diagnostic errors ranged from 5-9% and consisted of the lesions in Table III.

The major source of error shown in these series was differentiation from an adjacent neuroblastoma. From the imaging point of view this is certainly the main problem. However, when cases are collected from many sources, they must include non specialised centres. There will then be considerable variations in technique. The main imaging problem is to differentiate the intrarenal nature of a Wilms' tumour from the extrarenal compression of a neuroblastoma. The imaging errors are likely to arise from either using inadequate doses of contrast medium or a failure to take lateral radiographs. The problem in trying to analyse the work of others is that imaging today cannot just be accepted as a visual technique and the clinical parameters must be brought in before a pre-operative diagnosis is made. However, there is still likely to remain a small core

TABLE III

Diagnostic errors in Wilms' tumour

	Group I	Group II
No. of cases registered	606	442
No. of errors in diagnosis	30(5%)	40(9%)
Corrected diagnosis		
Neuroblastoma	9	19
Other cancer	7	7
Rhabdomyosarcoma	1	
Leiomyosarcoma	2	
Renal cell sarcoma	1	
Fibrosarcoma	1	
Lymphosarcoma	1	
Hypernephroma	1	
Cystic disease (includes polycystic)	9	11
Other	5	3
Adrenal hemorrhage	1	
Congenital renal thrombosis	1	
Dysplastic kidney	1	
Renal carbuncle	1	
Teratoma	1	

References:

Group I 1) D'Angio et al
 2) Ehrlich et al

Group II Lemerle et al

(1-2%) of problems in which even a combination of US and IVP fail to resolve. In these cases CT may prove the more effective answer.

Our current practise is to delegate the IVU into a secondary position compared to ultrasound. This will be further expounded under the ultrasound section.

Principle and Method

The IVU is the standard method for examining the urinary tract. It relies on an intravenous injection of Iodine containing contrast medium to outline the kidneys, ureters and bladder. A major disadvantage of the IVU is that severe reaction may occur following the injection of contrast medium and affect heart, kidney or lung as target organs. When serious, they may result in sudden death (Witten). This has resulted in recent moves to utilise either non-ionic or low ionic contrast media which have a lower toxic effect (Dawson & Grainger; Dawson et al).

Having noted this major disadvantage to the IVU, we must stress that it is not a problem in children. Children are very unlikely to have serious contrast reactions and over the last ten years with over 5,000 studies we have had only a few minor idiosyncracies of an anaphalactoid nature.

As children are not prone to serious contrast reaction we use a relatively high dose of contrast medium, i.e. in the region of 3-4ml of Conray 280 per

kilo. The normal dose—would be in the region of 2-4ml per kilo, but we use the maximum in an attempt to get consistently high quality IVUs when the problem of tumour evaluation is under consideration.

Technique

There is usually no need for any preparation of the patient. The following is our method (Aaronson & Cremin):-

- 1) Control film of the kidney, ureter and bladder area.
- 2) After the control film and injection, the child is given a fizzy drink to distend the stomach. This creates a gas window that displaces the colon and allows better visualisation of the kidneys, particularly on the left side.
- 3) A collimated "nephrogram" film at 3 minutes.
- 4) A full length "excretory" film at 10-15 minutes.
- 5) A lateral film (full length) is mandatory and is taken at 15 minutes.

The series consists of a minimum of four films and this may be sufficient for diagnosis. It is our duty as a radiologist to minimise the amount of ionising radiation that is used. However, this is a hazard that may be overemphasised, and when the question of the greater danger of cancer misdiagnosis arises, then we have no hesitation about taking extra films or tomograms as required.

A further point should also be made. At no stage do we use compression bands to enhance the image. The

rational behind compression is to hold up contrast medium excretion by compressing the ureters as they enter the bony pelvis. Compression is not only difficult to apply in children, but is contraindicated as it may cause tumour rupture.

Interpretation

The diagnostic features on I.V.U. have been well recorded (Dickey & Chandler; Hope et al; Lallie et al; Cope et al). The largest series was by Cope et al from Bristol, England. They studied the radiographs of 43 cases in whom 37 had an I.V.U. On the plain film, bulging of the flank was seen in three quarters of the cases, loss of renal outline occurred in two thirds and in the other third the renal outline was enlarged. Displacement of bowel gas was seen in half the cases, and the ipsilateral psoas shadow was obliterated in one third. Calcification was seen in 10% of tumours.

After injection of contrast medium, one fifth of the cases showed impaired renal function, two thirds showed displacement of the pelvicalyceal system and over one half some distortion. Enlargement varying from localised hydrocalycosis to dilation of all major calyces was demonstrated in nearly two thirds of the tumours.

These are the standard features to be looked for in Wilms' tumour. Our own experiences have been similar with the following comments.

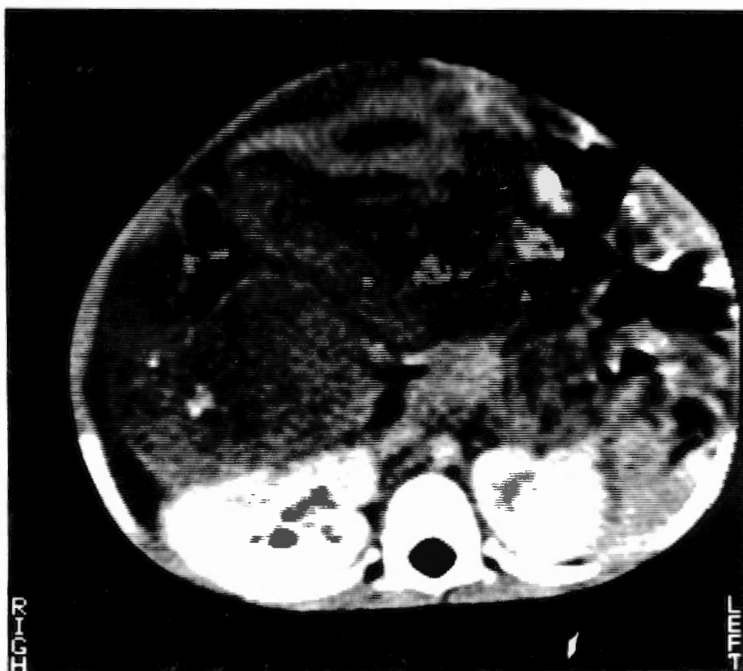
The control radiograph in the majority of cases will show an obvious fluid density mass so that observations on displacement of bowel gas or obliteration of psoas shadows are secondary or obvious features. Calcification was not a prominent finding occurring in about 5% of our cases, though this incidence may be increased three fold when C.T. is used (Fig. 2). On the plain radiographs this calcification may be streaky or irregular in form.

The crux of the whole matter of image interpretation is the demonstration of the intrarenal nature of the tumour. Most tumours are large so that in general this interpretation is not too difficult. An intrarenal mass occurs within the renal contour and causes both splaying and distortion of calyces. Elongation of the major calyces occurs with curvilinear displacement of them around the margins of the tumour. The minor calyces become flattened and increase in their transverse diameter. The overall picture is a bizarre but typical appearance. We have developed a three dimensional concept of visualizing the mass by utilising both frontal and lateral radiographs. It must be appreciated that outward expansion of the tumour is limited by the surrounding relatively rigid structures in upper pole and posterior tumours. At these sites the tumour will cause more calyceal distortion than lower pole or anterior tumours. Lower pole and anterior tumours more readily expand to the exterior and

FIG. 2



(a) Wilms' tumour of right kidney showing typical irregular calcification.



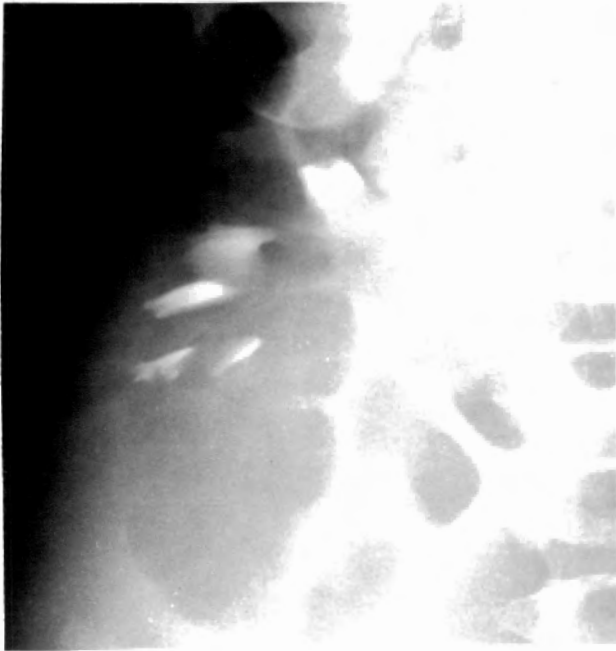
(b) CT of the same case showing the calcification in this anteriorly placed tumour.

therefore become exophytic (Fig.3). A centrally placed tumour may cause a hydronephrotic appearance associated with calyceal distortion.

Illustrations of upper pole and posterior tumours are shown in Figs. 4 and 5 and lower pole and anterior tumours in Figs. 6 and 7. The tumours often reach a large size and may cross over the midline. Large tumours may also virtually replace the excreting parenchyma to cause "non-function" or "non-visualisation" (Fig. 8). This may occur in between 5-10% of cases (Cremin) but does not appear to affect the prognosis (Canty et al).

The major problem as mentioned earlier is differentiation from an adjacent neuroblastoma that arises from suprarenal or sympathetic chain. The clinical appearance, lack of diffuse bony metastases and non elevated catechol amines are helpful. A neuroblastoma may produce both displacement of the kidney and pelvicalyceal system. Identification of displacement of the kidney identifies the diagnosis and is not usually difficult and U.S. or C.T. should resolve the problem. A Wilms' tumour does not usually extend across the midline in such a manner as to displace the kidney on the other side and this is also an important feature in differentiating it from a neuroblastoma. This point has been stressed by the radiologists from The Hospital for Sick Children, Great Ormond Street,

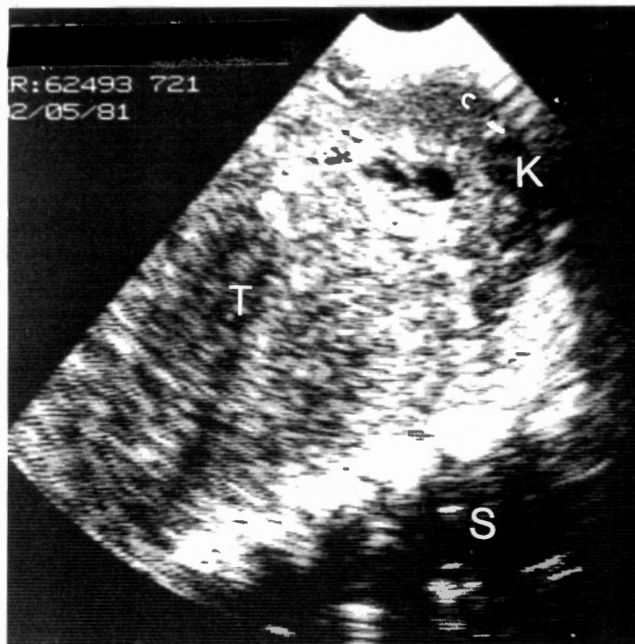
FIG. 4



(a) IWU upper pole tumour of the left kidney producing calyceal distortion.

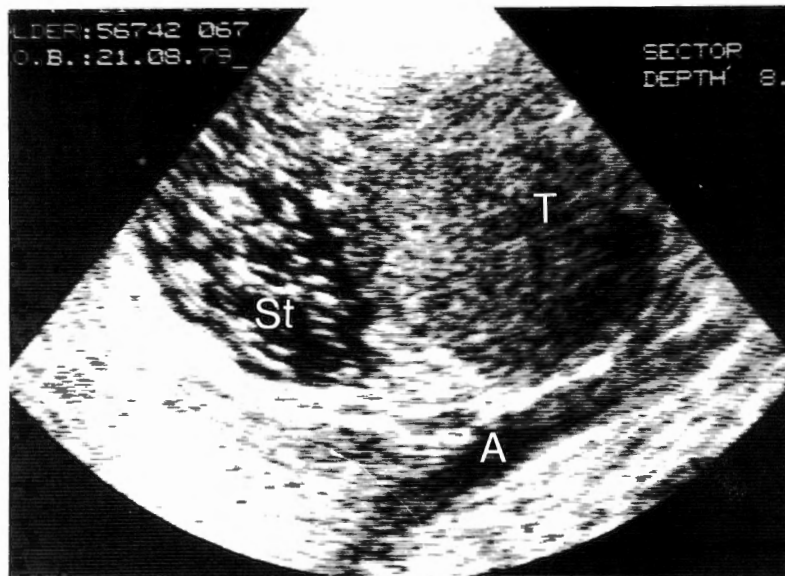
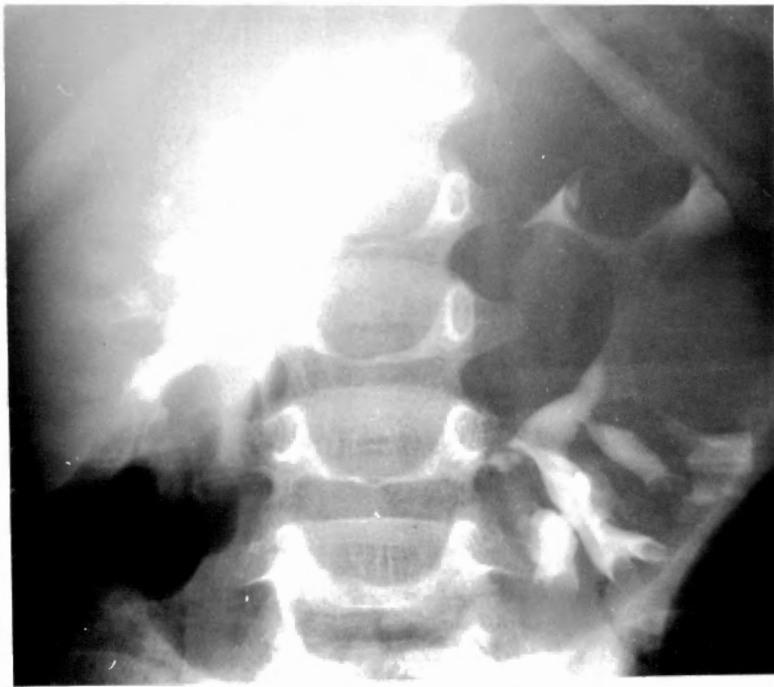


(b) Lateral projection confirming position.



(c) Sagittal US of the same case showing tumour (T) with areas of differing echogenicity but having an overall increased echogenicity when compared to kidney (K), dilated calyx (C), spine (S).

FIG. 5

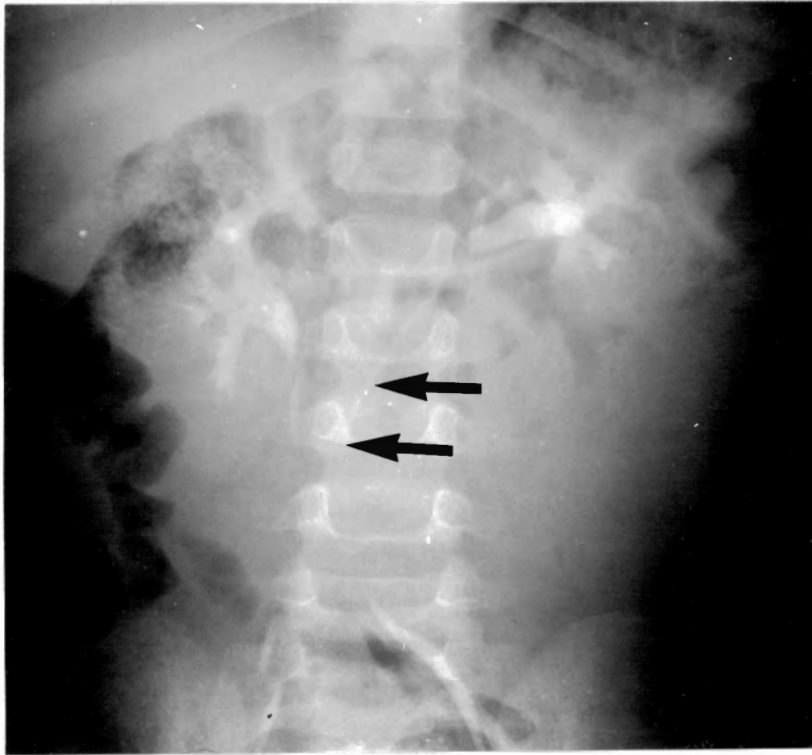


(a) IIV showing tumour in left kidney causing calyceal distortion.

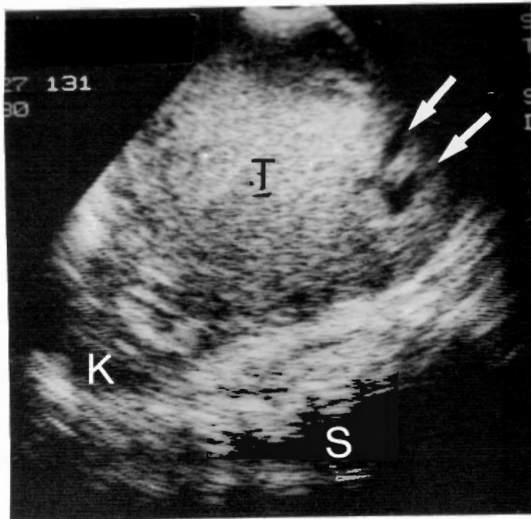
(b) Lateral radiograph shows the posterior position of the tumour (arrow).

(c) Coronal US showing its generally homogeneous nature. (A) aorta, (St) stomach, (T) tumour.

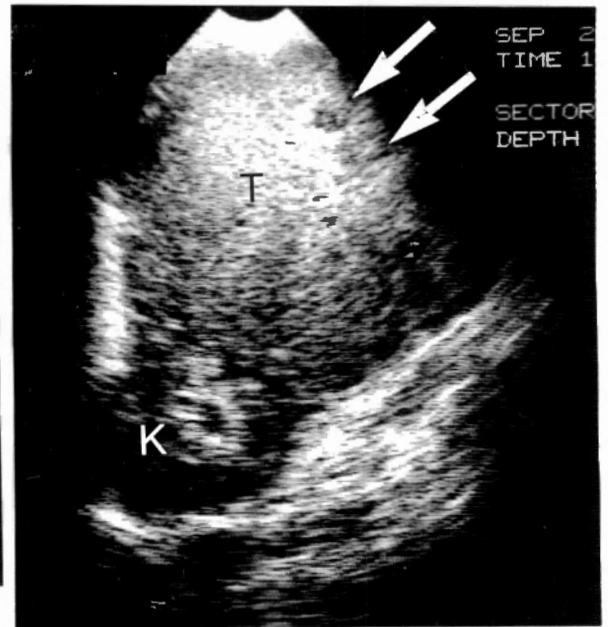
FIG. 6



(a) IWU showing massive tumour of the lower pole of the left kidney causing displacement of the ureter to the midline (arrows).



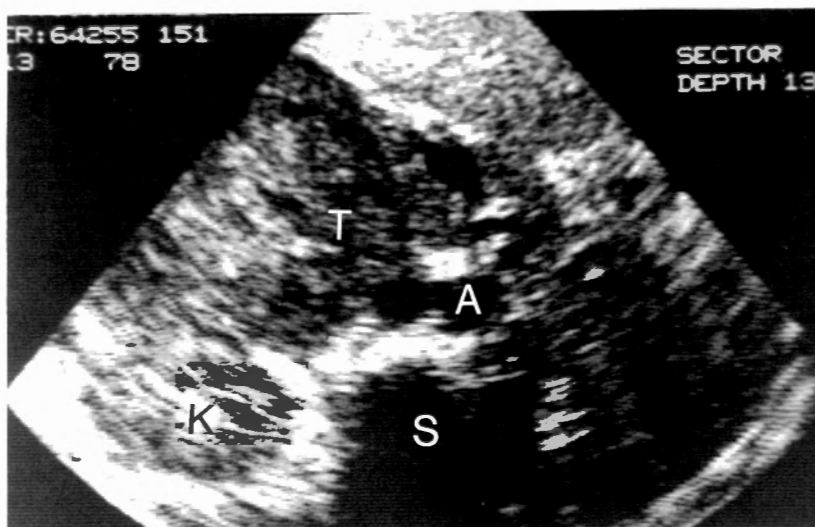
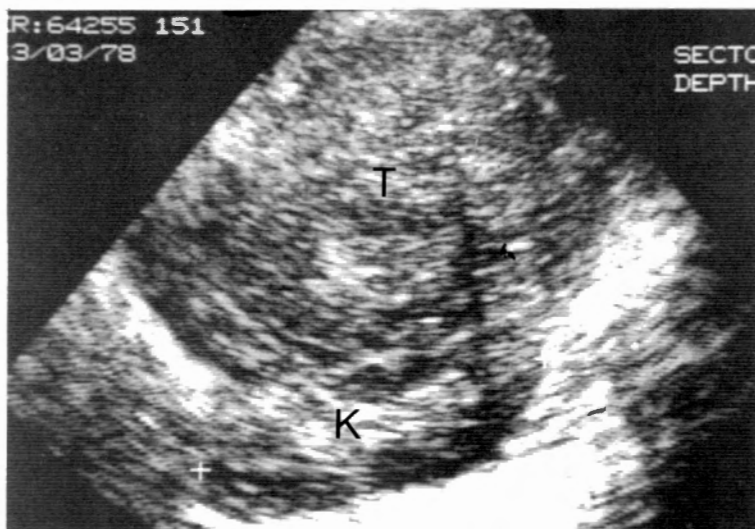
(b) Sagittal US showing tumour (T) arising from lower pole of kidney (K), spine (S). Break-down in tumour is shown by arrows.



(c) Transverse US showing that the tumour (T) is arising anteriorly.

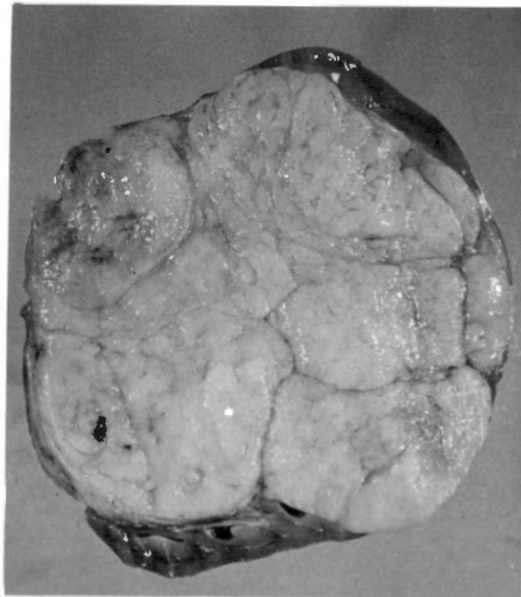
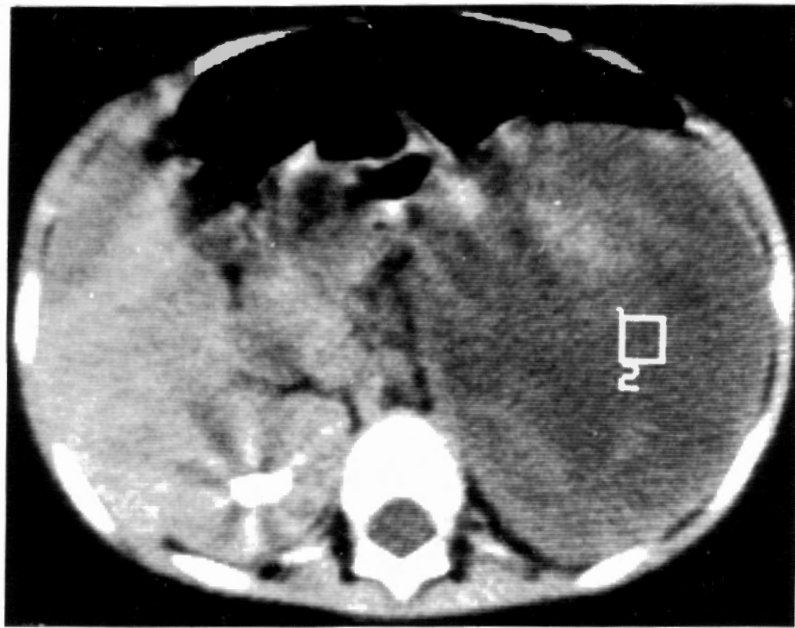
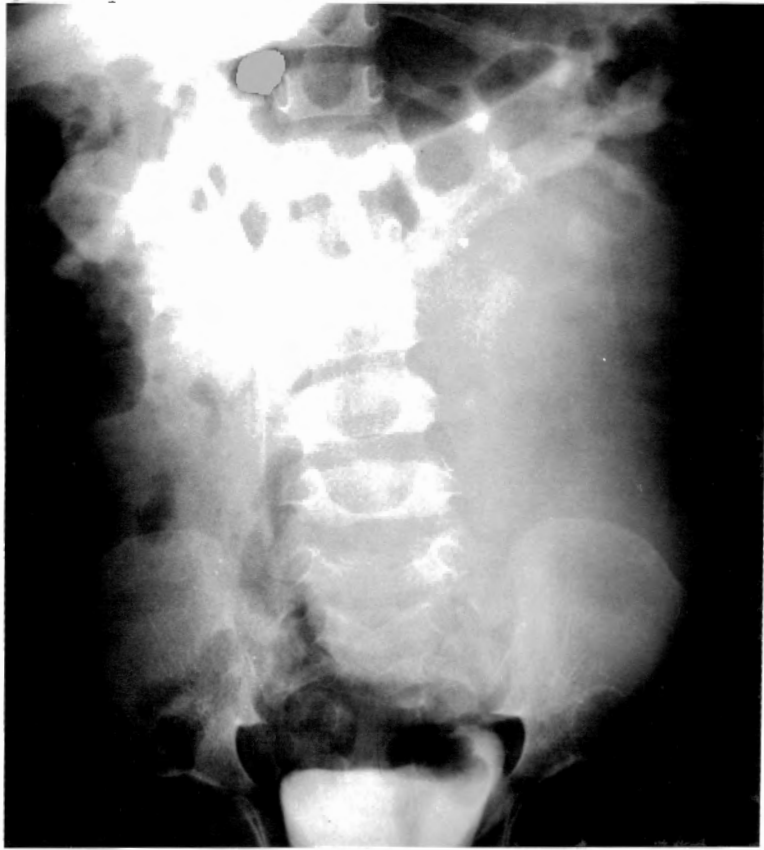
- (a) IVP showing mass in right kidney. There is some splaying of calyces but this is not gross.
- (b) Lateral radiograph of IVP. The renal mass is expanding in an anterior direction. As there is little resistance in this direction the mass does not produce marked calyceal splaying.
- (c) Sagittal US showing the anterior expansion of this tumour.
- (d) Transverse US showing the exophytic anterior position of this mass. The tumour is of mixed echogenicity and is extending medially over the spine. Tumour (T), kidney (K), aorta (A), spine (S).

FIG. 7



- (a) IIV showing "non-function of left kidney".
- (b) CT of same case showing replacement of kidney tissue by tumour mass. The cursor is in the centre of the tumour which had a lower density reading than the periphery of the tumour.
- (c) Postoperative specimen showing that the kidney has mainly been replaced by tumour so that only a thin rim of parenchyma persists at upper and lower poles.

FIG. 8



London, (Chrispin et al) and is undoubtedly true. However, there are exceptions and some of the tumours referred to us from outlying districts in Southern Africa have been enormous and have extended across the midline.

It must also be noted that several cases of Wilms' tumour have been reported following trauma (Persky & Forsythe; Fraley & Halverstadt). Pathological kidneys are particularly prone to injury, and this may follow minimal trauma (Slade et al). The diagnosis of Wilms' tumour should be considered when a grossly expanded kidney is seen on I.V.U. following a slight injury. Our own experience is that the trauma is the initial presenting feature in about 5% of Wilms' tumours.

ULTRASOUND

Ultrasound

The technical refinements of ultrasound in the last 10 years have completely changed our approach to abdominal masses in children. This has been an evolving process. Initially we had to transfer the patients to Groote Schuur Hospital for examination by a bistable unit that showed the solid nature of the mass in black and white. The detection of its intrarenal nature was difficult and the mass appeared as an echofree zone with occasional scattered short echoes within it (Hunig). Because of the lack of sensitivity in differentiation of relatively homogeneous solid masses which may produce no echoes from similar appearances in fluid collections we made our first error in 1976. This was when we thought a centrally placed echo free tumour might be a peripelvic cyst. The child had a Wilms' tumour and is still alive and currently attending the oncology clinic for routine check-up.

The next step in evolution was grey scaling in which it was easier to detect both the solid and the intrarenal nature of the mass. The apparatus was a B Scale unit with an articulated arm and considerable skill and patience was required to compound the images particularly when the child became restless.

A major advance occurred when the real time US scanners were developed. We obtained our first unit in situ installation in 1980. This was a mechanical Sector Scanner that had a small transducer head, ideal for

examining the child's abdomen. The unit produced grey scale computed images and it was relatively easy to visualise the kidney and acquire "frozen" pictures.

Principle and Method

At this stage a few physical properties will be mentioned. Ultrasound relies on production of an image by the reflection of high frequency sound waves from interfaces. The more solid, or to be more correct, the more cellular a structure, the more "interfaces" it has and therefore the more echogenic or reflective it becomes. The acquisition of grey scale images in which some tonality is given to the reflection is a product of prolonged exposure photography. The term real time arises from the computer world and refers to the capability of the computation to keep up with the process it is measuring or controlling (Winsberg). With its advent it became possible to image moving structures continuously within the body. The real time units have a digital memory (scan converter) that can be frozen to produce an image that is compatible with television and multiformat cameras. This technology has transformed the imaging of the paediatric urinary tract.

Before the advent of real time units it was our custom to first examine the child with an I.V.U. and then to confirm the findings by U.S. The I.V.U. gave an initial map and it was then possible to focus on relevant pathology by US. A disadvantage was that the

carbonated drink in ~~the~~ stomach which provided an air window for the IVU which obscured the sonic images from the left kidney.

When real time ultrasound became available the examination, though still requiring considerable skill, became much easier and with practice we were able to confidently predict our findings without initial localisation by an IVU. The current status is that ultrasound is now the primary method of examination with the IVU delegated to a secondary position.

The current objectives of our ultrasonographer when presented with a suspected renal mass are:-

- a) To confirm site of lesion, i.e. in the kidney.
- b) To ascertain its gross acoustic morphology.
- c) To exclude local invasion, e.g. into vena cava.
- d) To exclude distant metastasis, e.g. into liver.
- e) To confirm the contralateral kidney is normal.

Technique

The child initially lies supine on the examination table. No sedation is usually required and the child is distracted by the familiar television screen. A water gel interface is applied to the child's abdomen before contact is made with the transducer.

The examination must be carried out in sagittal, parasagittal, coronal and transverse planes (Cremin). The examination of the right kidney is relatively easy, as in the supine position the liver can be used as an acoustic window. When the left kidney is examined a

coronal approach makes use of the spleen in a similar fashion. In most cases the examination can be completed from the supine position but if there is any problem from bowel gas interposition, the examination is completed from the lateral decubitus or prone positions.

Appearances

An intrarenal tumour usually bulges beyond the kidney margins. It is predominantly solid, slightly hyperechoic (more echogenic than adjacent liver) and causes gross renal enlargement. A clear margination of the tumour from the rest of the kidney may be provided by a pseudo capsule (Jaffe et al) and this may give a border that is echo free or echo dense. Alternatively the margin may be an abrupt change into normal kidney texture. Distorted, obstructed or displaced calyces may be seen within the kidney as sharply marginated hypoechoic areas. The overall appearance may appear homogenous (De Campo) but small areas of increased echogenicity usually exist (Fig. 4c,7c). This often becomes more apparent on altering the gain factors. Highly hyperechoic foci causing refraction and acoustic shadows indicate calcification, whilst large hypoechoic areas may indicate foci of haemorrhage. These areas become necrotic and liquify so that they become hypoechoic (Fig. 6b,c). Localised areas of necrosis are fairly commonly seen on US. (Gates et al) and in this respect US can be more sensitive than CT in which

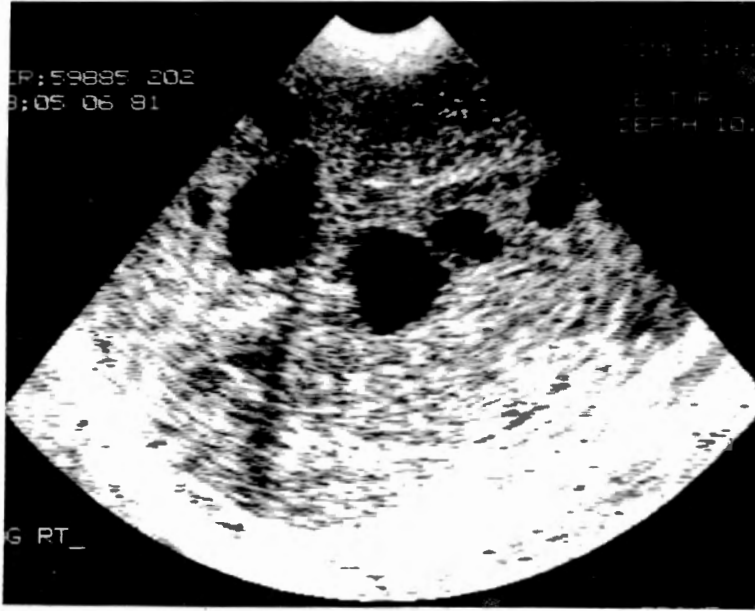
necrosis shows as areas of lower attenuation (Fig. 9). In multilocular cystic nephroma (Wood et al) a more florid cystic appearance is seen with lakes of fluid and floating septae having a fucoid appearance (Fig.10). Fluid can cause acoustic enhancement so that parts of the lesion may appear more solid than they are.

Ultrasound can detect invasion of the vena cava (Slovis et al, 1981) and extension into the right heart (Slovis et al, 1978). It may be superior to venography (Fig. 11) as the variations in diameter of the vena cava that occur with posture and respiration can be readily detected by real time units. We have been successful in demonstrating invasion of the IVC by US in only one case during the last 5 years (Fig. 12).

An ultrasonic diagnosis of synchronous bilateral tumours has been made once in our last 27 cases. There were no special ultrasonic features and the diagnosis was confirmed by other imaging techniques Fig. 13.

We have not been able to identify ultrasonically any localised areas of nephroblastomatosis. Montgomery et al have reported that the focal lesions shown by CT (Fig. 14) may be normal on US. Their recommendation is that the best method of imaging nephroblastomatosis is by CT. We have no case to show and have therefore included Montgomery's CT case in this section. We also have included another of his cases to show that CT is better at differentiating small cysts which might be considered focal areas of nephroblastomatosis on US

FIG. 9



(a) Sagittal US of right kidney showing cystic necrosis in tumour.

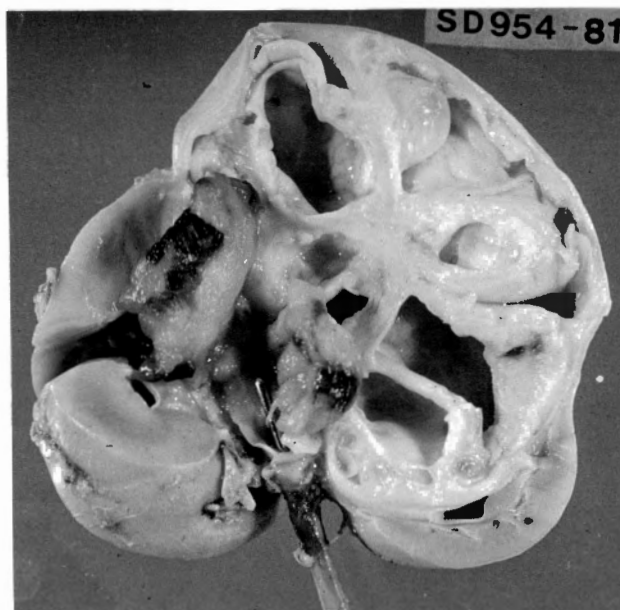


(b) CT of same case shows corresponding areas of reduced attenuation in the kidney. At surgery large areas of "fluid" were seen in the kidney.

FIG. 10

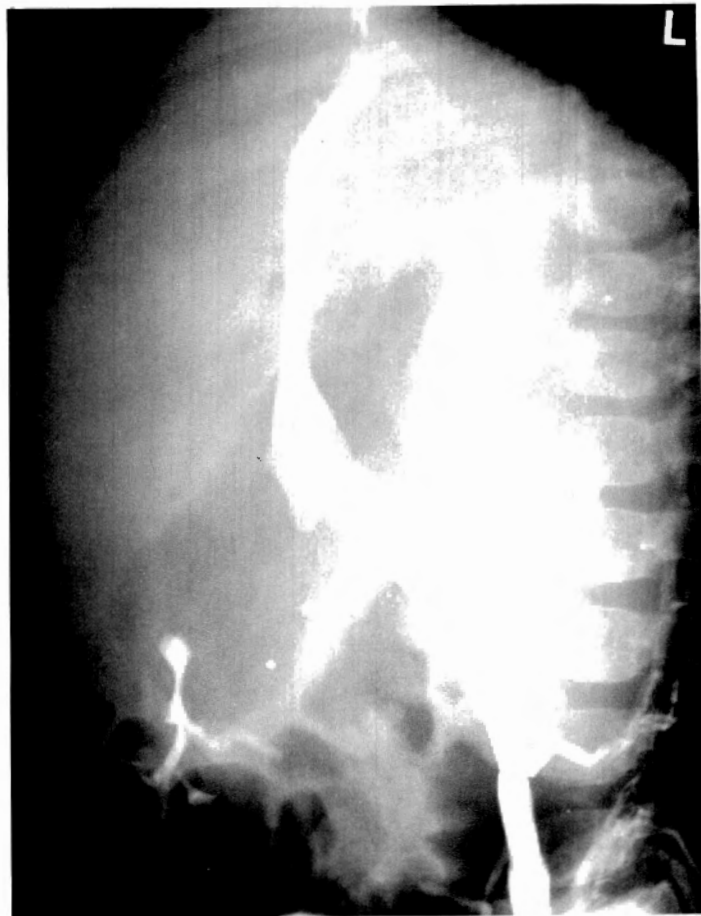
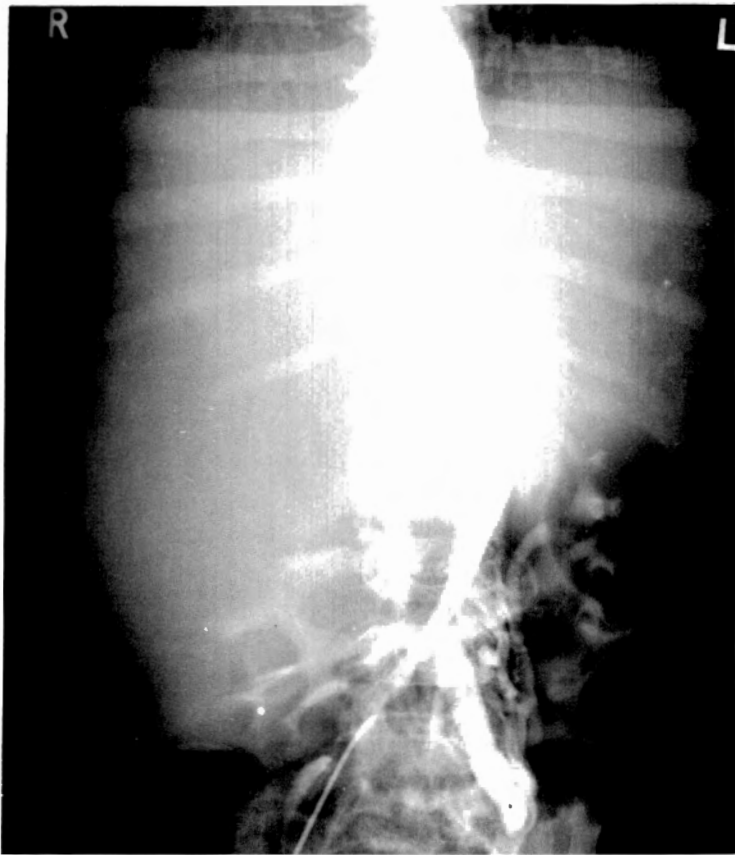


(a) & (b) Transverse and sagittal US of multicystic type of Wilms' tumour.



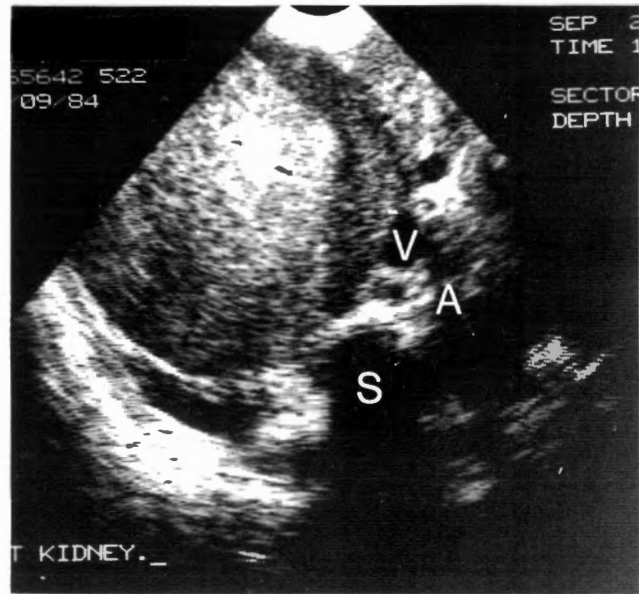
(c) Postoperative specimen showing the cystic nature of the mass.

FIG. 11

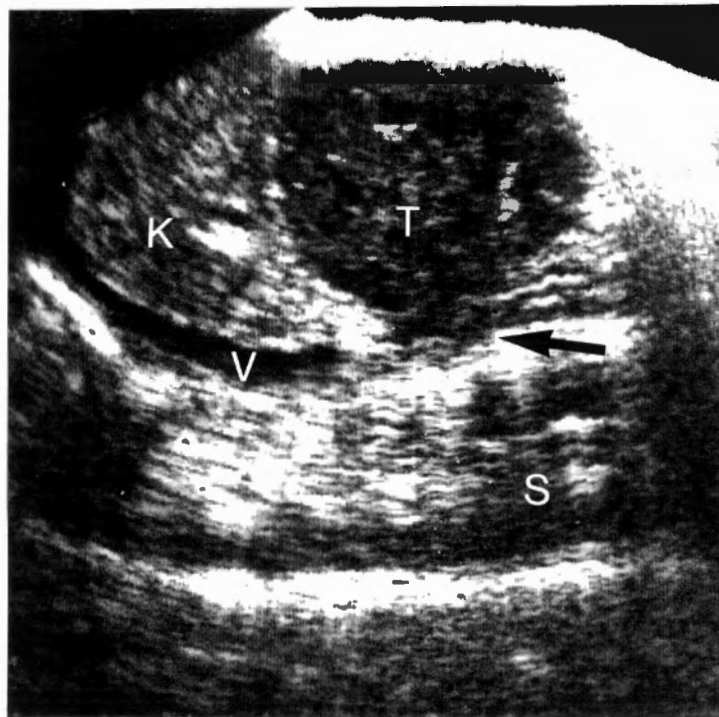


(a) & (b) Inferior vena cavagrams. Injection of contrast media via a catheter from the femoral vein and iliac vein. The inferior vena cava shows medial and anterior displacement by the tumour but no invasion.

FIG. 12



- (a) Transverse US showing massive tumour in right kidney displacing the vena cava (V). Aorta (A), spine (S).



- (b) Sagittal US. The arrows show lower pole tumour (T) invading vena cava (V). Kidney (K), spine (S).

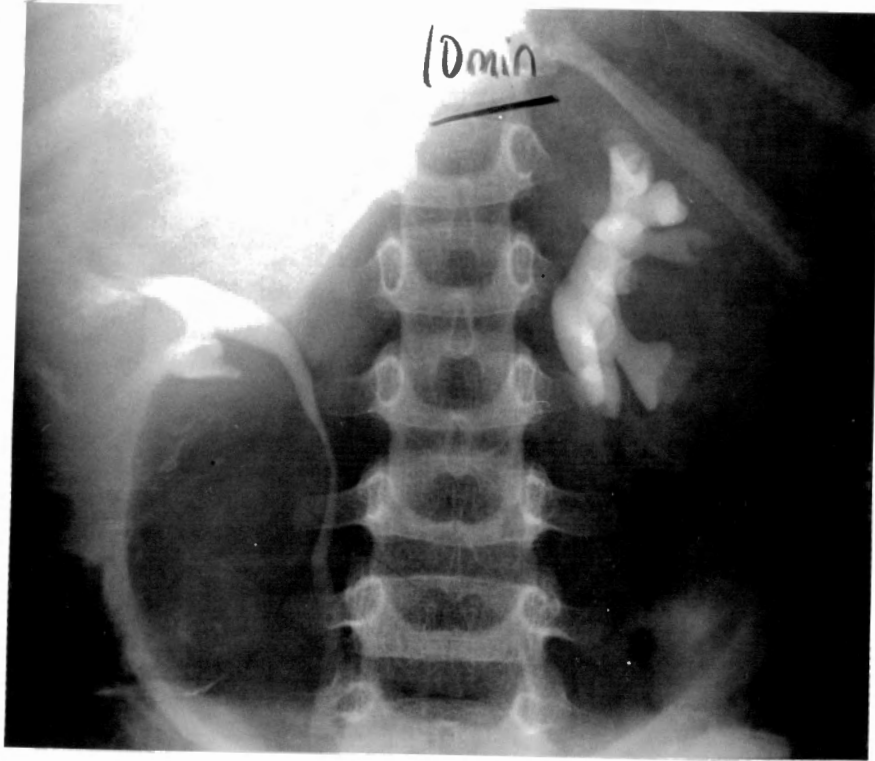
(a) IWJ at the 10 minute phase. There are bilateral Wilms' tumours that are causing pelvicalyceal displacement. On the left this displacement is upwards and medial and could be mistaken for a malrotated kidney.

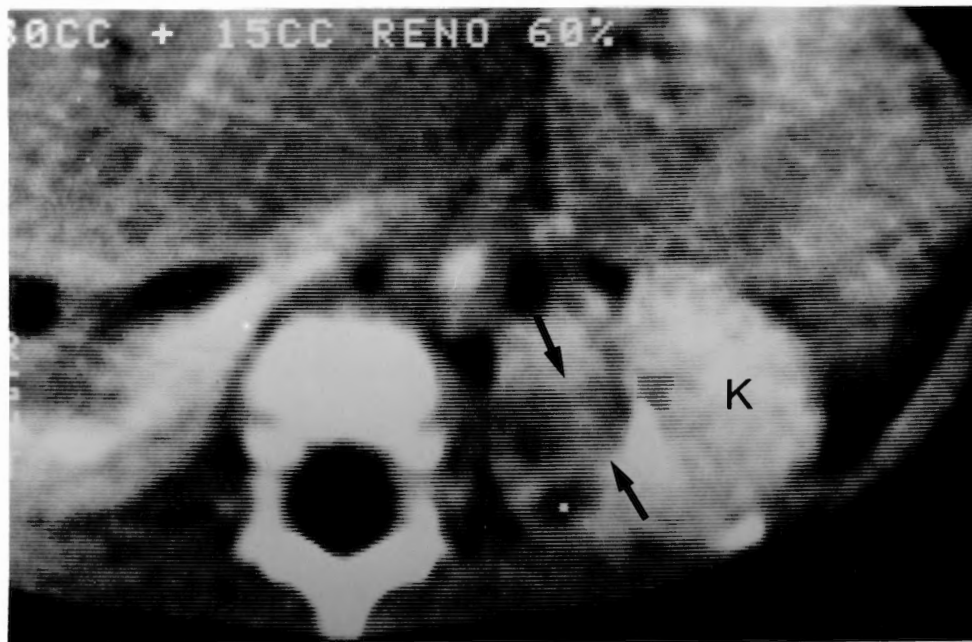
(b) The true situation is revealed by tomography in which a mass is shown in the lower pole of the left kidney.

(c) CT of same case showing an anteriorly placed right tumour and a posteriorly placed left tumour. There is some contrast enhancement of kidney parenchyma at the rim of the tumour.

Comment: As is to be expected, the posteriorly situated tumour is causing more pelvicalyceal displacement.

FIG. 13





This patient had a Wilms' tumour of his right kidney. The CT scan of the left kidney (K) showed two discreet low density lesions (arrows). Wilms' tumourlet was identified at surgery.

Acknowledgement to P. Montgomery et al, Pediatric Radiology 1984 14:392.

(Fig. 15). We conclude the section with the belief that the only method of excluding nephroblastomatosis is by palpation and biopsy at the time of surgery.

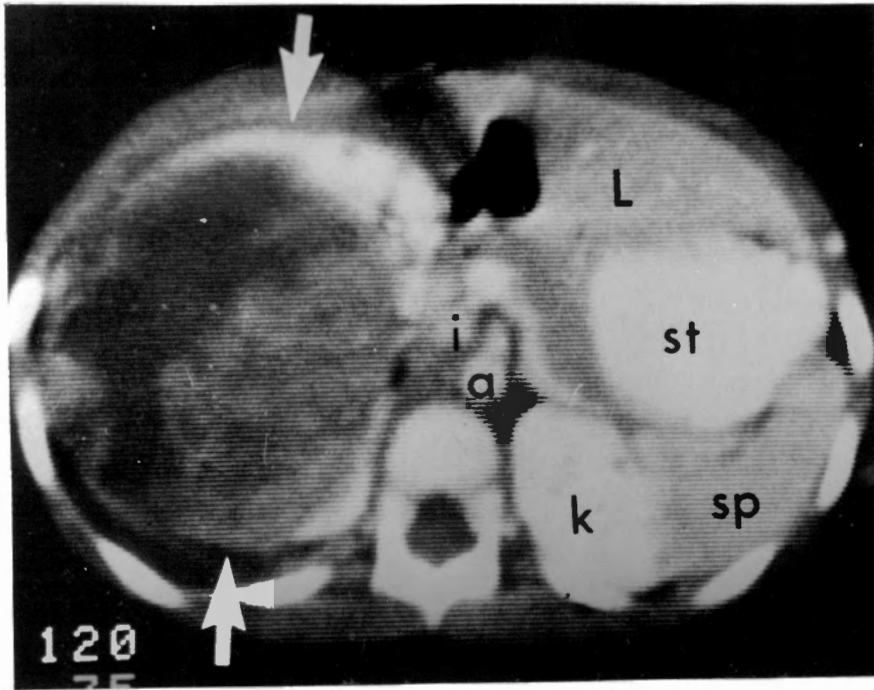
The above has been a description in general terms. However a more detailed survey of our last 27 cases during the 5 year period (1981-85) showed that we were able to make, by ultrasound alone, a confident and definitive diagnosis of Wilms' tumour in 18 cases.

In the other nine cases we strongly suspected Wilms' tumour but preferred to have a second examination such as an IVU or CT to confirm the diagnosis.

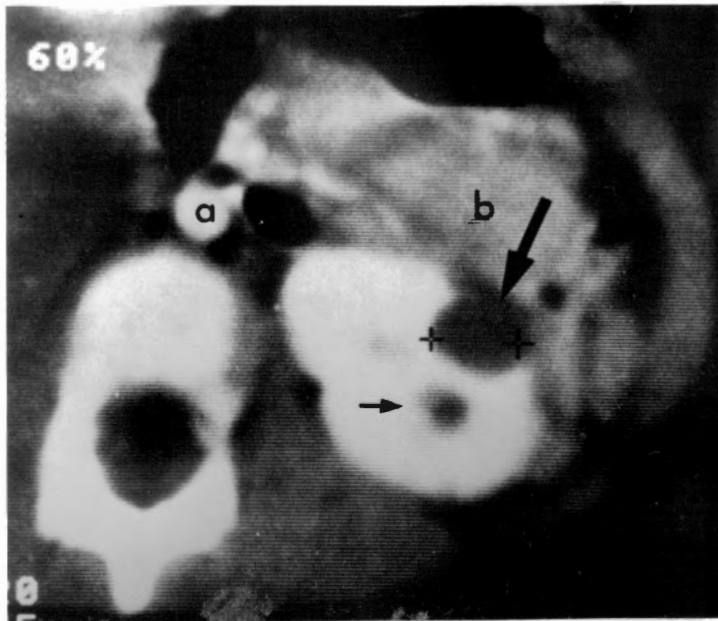
The US findings (Cremin) in these 27 cases were:-

- 1) All were tumours 5-10cms in diameter.
- 2) None were completely homogeneous, although 5 cases had a superficially uniform appearance that could be altered by changing gain factors.
- 3) 22 cases had multiple 3cm areas of differing echogenicity.
- 4) 6 had fluid areas denoting necrosis or haemorrhage.
- 5) 3 had punctate areas of echogenicity that cast acoustic shadows and were confirmed to be areas of calcification.

On reviewing the literature on the ultrasonic detection of Wilms' tumour, we find there is no great depth of publications. There are relatively few articles dealing with the grey scale characteristics of Wilms tumour and except for publications by De Campo



(a) Wilms' tumour (arrows) seen on the right. Vena cava (i), aorta (a), liver L, stomach (st), spleen (sp), left kidney (k).



(b) CT scan of the left kidney shows two innocent cortical cysts. Aorta (a), bowel (b), cysts in left kidneys (arrows).

Acknowledgement to P. Montgomery et al, Pediatric Radiology 1984 14:392.

they only deal with a limited amount of cases (Cohn et al; Hartman & Sanders; Jaffe et al; Kuhn & Berger; Mulhern et al). One of the earliest real time accounts was by Jaffe et al who reported a spectrum of sonographic appearances in 11 cases. Most tumours were large, predominantly solid and well circumscribed. None were purely cystic, small anechoic areas were seen in most cases and were related to small areas of haemorrhage or necrosis within the tumour. The account by Hartman & Sanders from the Armed Forces Institute of Pathology, Washington D.C. retrospectively reviewed the sonograms of 46 cases of Wilms' tumour and neuroblastoma. A correct diagnosis was made in 23 out of 26 "unknown" cases of Wilms' tumour and neuroblastoma on the basis of the sonographic findings alone. The series included one case of Wilms' tumour arising outside the kidney and one case of neuroblastoma invading the kidney.

The most recent account is by De Campo from the Royal Children's Hospital, Melbourne and is the largest published series, consisting of 30 cases. It is not possible to compare our results directly with theirs as they were performed on an "Octoson" Water Delay Scanner. This scanner uses 8 transducers and a water bed so that complete transaxial panoramic images in multiple planes are produced. The findings were essentially similar to ours and a confident diagnosis of intrarenal tumour was made in 21 cases. In the other 9 cases the ipsilateral

kidney could not be distinguished from tumour. In these cases the question of neuroblastoma arose and an IVP was performed to confirm the diagnosis. They were also reluctant to diagnose Wilms' tumour on US alone when the lesion had a cystic pattern. 87% of the cases showed the mass to have a uniform or homogeneous texture. In this respect our findings differed. The explanation for this is likely to be twofold. Firstly the "Octoson" produced a very much more smoothed-out and uniform image to the ATL Mechanical Sector Scanner that we employ. Secondly, our interpretation of homogeneous was different from theirs. The tumour may look superficially uniform but careful inspection will show small areas of different echogenicity. In their series unless these areas were greater than 1cm they were called homogeneous. No liver or nodal metastasis were picked up and in 27 cases the inferior vena cava was patent. The interpretation in the other cases was inconclusive but no thrombus was found at surgery.

There has also been another recent ultrasound report from Australia (Morrison). This concerned the detection of Wilms' tumour in 3 adult patients (17 years, 21 years and 33 years). The findings were similar to those seen in children and consisted of hyperechoic tumours in the kidney. The appearances on their own were not distinctive and would not help to differentiate the tumours from renal adenocarcinoma.

COMPUTED TOMOGRAPHY

Computed Tomography (CT)

The extent and nature of the tumour is well demonstrated by CT. However, young children lack the abdominal fat that produces the dramatic CT images seen in adults. To acquire satisfactory images needs a careful technique, a fast scanner (below 2secs), adequate sedation and contrast enhancement.

The early reports on CT that used scan speeds of up to two minutes are not comparable to current results. Leonides et al from Tufts New England Medical Centre compared CT and IVU. There were only two cases of Wilms' tumour and in one of them both CT and US showed improved visualisation compared to IVU. Damgaard-Pedersen et al from Copenhagen compared CT with RNS. They only had one case of Wilms' tumour and this was better shown by CT than RNS.

In 1981 a whole volume of the Radiological Clinics of North America was devoted to Pediatric Computed Tomography and two articles by Kuhns and Kuhn & Berger were devoted to CT of kidney masses in children. The latter article was from Buffalo Children's Hospital, New York and noted that CT had been used in their last 16 cases of Wilms' tumour. They further stated that if they had relied on IVU they would have had two errors of diagnosis and that there would have also been three cases in which the diagnosis was uncertain. Ultrasound improved their diagnostic accuracy to one error and two equivocal cases. No details of these cases were given

but it was stated that CT was one hundred per cent correct. We do not debate the advantages of CT and would also use it routinely if we had it on site. However, some comment is necessary on the cases in which US was stated to be equivocal. US requires skill and practice and in 1981 we would also have had doubtful or equivocal cases. However now that we have had some 5-6 years concentrated experience using real time scanning as a primary method of examination very few cases remain dubious after a combined US and IVU examination are made. Diagnostic problems with any form of imaging usually involve differentiation of nephroblastoma from neuroblastoma and Kuhn and Berger give a useful diagnostic tabulation (Table 4).

Principles and Methods

In CT a narrow beam of x-rays is passed through the child and the amount of transmitted radiation is detected and recorded by an array of crystalline detectors placed opposite the source. The x-ray source moves around the patient and a large number of individual measurements are made during this rotation. The measurements of differential x-ray absorption are recorded in a computer and used in a series of complex mathematical calculations to determine the radio-opacity for each square centimeter of body area. This digital information is then used to build an axial transverse scan section of the body. The advantage of this method

TABLE IV

Characteristics of Mass	LOCATION OF MASS	
	Intrarenal (Splays Kidney)	Extrarenal (Displaces or may invade kidney)
Shape	Round	Irregular, occasionally round
Margins	Tend to be regular; may have "pseudocapsule"	More likely to be irregular, lobulated; well-defined "capsule" occurs infrequently
Center	Mixed, low-density; 25-30% show slight calcification	Mixed low-density; 75% show some calcification, often extensive
Extension Vascular	Displaces vena cava & aorta; may grow into vena cava	Displaces, surrounds & encases vessels
Midline	May cross midline, but does not extend prevertebrally	Prevertebral midline extension is common

Reference: Kunn & Berger

of imaging over conventional radiology is its greater sensitivity. It will detect density differences of as little as 0.5% compared to 4-5% for the usual radiograph. It becomes possible to visualise internal structures of the body that show no detectable differences in density on the ordinary radiograph.

However it must be remembered that our visualisation of the urinary tract by conventional radiology relies on opacification of the urinary tract by contrast media so that some of these inherent advantages of CT are less obvious. Nevertheless the advent of the latest generation of sub 2 second scanners has made great advances in the art of urinary tract imaging.

CT technique

A careful pre scan protocol is essential for a successful examination. It cannot be over-emphasised that the radiologist concerned must personally supervise all stages and not delegate these preliminaries. Nothing is a greater waste than to transport a child to another hospital, spend an hour of valuable scanning time and be unable to produce satisfactory images because veins cannot be found or the child is inadequately sedated. We have established the following routine (Cremin & Mervis) and consider it important enough to describe in some detail.

1. Patient evaluation

Paediatric CT body scanning is performed one

afternoon a week at the Groote Schuur Hospital. In the morning the radiologist visits the child in the ward. Familiarisation is important and the procedure should be explained to both the child and parents (who often attend the examination). The sites of suitable veins for injection are now ascertained and the child checked for weight, allergies, or any medication problems. We used to insert an intravenous line into the upper limb at this stage, to diminish the risk of disturbing the child with an injection just prior to the scan. We have abandoned this as the drip often malfunctions during ambulance transport. The intravenous injection can be given on the CT scanning table without unduly disturbing the child provided a suitable vein has been located beforehand.

2. Sedation

Vallergan and Droperidol are given orally at midday. The dosage is Vallergan 4mg/kilo and Droperidol 0.2mg/kilo. Preference is for administration by a nasogastric tube. There are two reasons for this, (a) the correct dosage reaches the stomach without being spat out or vomited up. (Knowing this is reassuring if the child gets initially restless when placed on the CT table). (b) Relatively large fluid volumes of contrast media can also be administered at the same time.

3. Contrast Administration

Bowel contrast enhancement is with 3%

gastrograffin.

Age ---	Gastrograffin -----	Water -----
6 months - 1 year	6ml	180ml
3 years	8ml	270ml
6 years	12ml	360ml

If necessary the bowel contrast media can be topped up by another 50ml of the 3% gastrograffin injection down the tube immediately prior to the examination. This is not usually required with Wilms' tumour. However when other paravertabral masses such as neuroblastoma or glands are to be considered then the duodenal loop should be opacified to act as a landmark in their evaluation.

On arrival at the CT room the following takes place:

- a) The child is given an IV contrast medium injection (2ml per kilo) and the bowel 3% gastrograffin topped up if required. We used to give 1ml of buscopan IV (hyoscine butylbromide) to reduce bowel motility but with sub 5sec scanning speeds this is not necessary.
- b) The child is immobilised by wrapping in a blanket and lightly strapped to the scanning board. The arms are kept extended above the head and sandbags may be used for immobilising the legs.
- c) Image sequence The procedure is to scan from xiphoid to umbilicus (lower limit depends on size of mass) using 1cm slice widths.

There may be variations on this, to include an

initial scan without contrast medium, or the use of a bolus injection.

For this technique a large gauge needle (19g-21g) is placed into the upper extremity prior to the scan. A relatively large dose of contrast medium (2ml of Conray 280 per kilo up to 100ml) is divided into two syringes. The first syringe is injected rapidly and after the injection scanning is commenced. The second syringe is then immediately injected and scanning continued during the injection.

The technique delineates the anatomy and relationship of pathology to major vascular structures. It may also improve definition of the mass and improve its image quality.

CT appearances

CT will confirm the presence of an intrarenal mass and demonstrate the characteristics of a Wilms' tumour.

Our experience since 1979 covers 14 Wilms' tumours and we have found these characteristics to be present:-

1) A tumour mass that distorts the renal outline and parenchyma. The mass is usually less dense than renal tissue but may be isodense in many areas.

2) The tumour is usually large and when it arises from the upper pole of the right kidney may cause apparent hepatic filling defects. These low density areas blend with the liver substance to mimic hepatic masses. This is later illustrated in Figure 23.

3) After injection of contrast medium the intrarenal nature becomes more apparent and the calyceal system is seen to be displaced. As with the IVU the amount of distortion and displacement depends on the site of the tumour and it will be less when the tumour is predominantly exophytic and anterior (Fig. 13).

4) Some contrast enhancement of the tumour occurs but this may be neither marked nor uniform and tends to affect the peripheral more than the central portions of the tumour (Fig. 8).

5) Areas of nonenhancement occur which may signify areas of old haemorrhage or necrosis as they tend to correspond to the anechoic areas seen on US (Fig. 9).

There is usually no difficulty in diagnosis and normal variants that may present as renal masses should cause no problems. These include the renal hilar lips that are formed by an infolding of the renal cortical substance at the renal sinus. This may simulate a mass on conventional nephrotomography. At CT the appearance of the kidney and early cortical contrast enhancement clearly shows the normal relationship. The same principle applies to pseudotumours formed by the Columns of Bertin or by regenerating renal nodules in reflux nephropathy. These will all show contrast enhancement identical to the normal kidney. Sinus lipomatosis and parapelvic cysts are not problems in the paediatric age group but CT will rule out these possibilities in the older patient.

The contralateral kidney must be examined carefully for evidence of a synchronous tumour. Areas of non-enhancement with different areas of attenuation must be interpreted with caution but nephroblastomatosis should be considered (Fig. 14).

The patency of the IVC may be defined by using a bolus injection which is combined with dynamic scanning through the liver or at the upper pole of the right kidney (Fig. 15). The upper limb is recommended for the injection as fewer artefacts may occur (Glazer et al, Barnes et al). Our experience with this technique is limited to only a few cases all with negative results. The reported findings (Marks et al) are enlargement of calibre of IVC with identification of a thrombus filling defect inside the contrast filled IVC. The same problems that are seen in cavography with flow related artefacts mimicing a thrombus may also arise.

As regards spread of tumour, CT may demonstrate involvement of adjacent lymph nodes that displace the contrast filled duodenum. Interpretation of paraortic and pericaval soft tissue areas is not easy and will require confirmation by surgical sampling of lymph glands for staging purposes. CT is also an excellent modality for detecting liver and lung metastasis and this will be further discussed in the follow up protocol on page 101.

Radiation dose

It will be appropriate to end this section on CT by making some reference to the amount of radiation involved in a paediatric CT examination.

The CT unit that we are currently using is an Elscint 1800 model that is capable of sub 2 sec scanning speed.

To assess the radiation dose received by our children this unit must be compared to other similar CT scanners. The General Electric 8800 and the Siemens Somaton II are comparable units and figures of radiation dose for paediatric use have been published by Kirks. They were 2-3 rads skin dose for each complete abdominal or chest examination. In order to express this in terms comparable to conventional radiology, this is the same dosage that is acquired by 3 minutes of fluoroscopy (Poznanski). The amount of fluoroscopy that we would use in a routine barium study would usually be about 5 minutes. We may therefore consider that the radiation dose received by the child in two CT scans would be about the same as one barium study. We believe this to be a totally acceptable figure.

MAGNETIC RESONANCE IMAGING

Magnetic Resonance Imaging (MRI)

MRI has recently been advocated as a major method for evaluating abdominal cancer in children (Cohen et al). To date there has been only one report (Belt et al) that covers its use in Wilms' tumour. I have not yet seen this publication but the second author Cohen is an ex South African who has written a book on Pediatric MRI. He has corresponded with me and kindly sent the manuscript of the article. The material is from the Riley Hospital for Children, Indiana, and covers 14 cases. It concludes that MRI has the potential for providing the same information as Computed Tomography, Ultrasound, liver spleen isotope scans, and excretory urography. Although expensive it can be cost effective if and when it replaces all of the above modalities!! What does emerge from this report is that although MRI is effective for imaging the primary tumour it gives no accurate prediction of capsule invasion. It does not (as hoped) affect the early staging of the tumour. Capsule invasion was predicted in four of the cases but was not proved correct by biopsy samples. MRI was reported to give excellent visualisation of the vena cava. This was involved in 2 of the 14 cases but this extension was also predicted by ultrasound. In the detection of liver and lung metastasis MRI was inferior to CT. On T1 weighted spin echo images the metastases were isointense with liver and difficult to detect. They were best seen on T2 weighted spin echo sequences

when they were of greater intensity. On inversion recovery sequences they appeared to be of lower intensity. The acquisition of all this data to form T1, T2 and inversion recovery images is time consuming and currently offers no advantage over CT imaging.

Principles and Method

We have had access to the Elscint Gyrix 0.5 Tesla unit at Tygerberg Medical Research Unit for the last six months.

Some understanding of the basic principles underlying tissue magnetization is necessary before showing some examples of its use.

The two forms of energy involved in creating MR images are radiowaves and magnetism, both of which surround us continuously in our natural environment. The use of these forms of energy (radiation) has not been reported to have any deleterious effects in the human body (Cohen).

In the following few pages an account of MR imaging will be given. Much of it has been taken from a lecture by Newhouse on the subject at the Society of Uroradiology meeting in the Hague in May 1986.

When the body part to be imaged is placed in a strong magnetic field, many of the atomic nuclei tend to align magnetically in the same direction as the magnetic field. This is known as longitudinal magnetization. If, in addition to the main magnetic field, the tissue is

subjected to pulses of radiofrequency energy, the tissue's magnetization can be made to spin in a plane perpendicular to the main magnetization (the transverse plane). This rotating magnetization of the tissue causes the tissue to emit a radiofrequency signal of its own. This emitted signal is detected and used to create the MR image. The intensity of the image which is created is directly proportional to the strength of this signal, which in turn is directly proportional to the strength of the magnetization of that tissue.

In practical terms, the nuclei which create the tissue magnetization are protons, the nuclei of hydrogen atoms. These are primarily in water and lipids, so that the images are primarily of tissue, water and fat. Some of the tissues in the body have relatively few of these protons which are susceptible to magnetization (e.g. dense fibrous tissue and cortical bone); these regions, therefore, are unable to contribute to the image and appear black. Moving tissues such as blood emit a signal which diminishes extremely rapidly, so that large blood vessels also appear black.

T1 (longitudinal) relaxation is a phenomenon which occurs to different degrees in different tissues, and can be made to provide tissue contrast. In brief, T1 relaxation is the process by which the magnetization of the tissue parallel to the main magnetic field increases to reach its maximum value. This longitudinal magnetization can be destroyed by each of a series of

90° pulses; the rate by which this recovers between the pulses varies from tissue to tissue and the amount of recovery of this magnetization depends upon the time interval between the pulses. The time interval between successive 90° pulses is known as TR; when it is relatively short (a few hundred milliseconds), the resulting picture shows a great deal of gray-scale variation determined by variations in T1 among tissues; when TR is long (a second or two), there is little or no T1 variation in the resulting images.

In general, the more "watery" a tissue is, the longer its T1, and the darker it appears in T1-sensitive images. In the urinary tract, the medulla of the kidney can be made to appear darker than the cortex due to the medulla's longer T1. Renal cysts may appear darker than any solid tissue for the same reason. Urine in the hollow viscera (which, like cyst fluid, has a very long T1) can be made to appear considerably darker than the walls of the structures containing it. Aqueous tissues (such as kidney, adrenal, solid tumours, bladder wall, etc.) will all appear to be darker than fat (which has a very short T1).

T2 (transverse) relaxation is the process by which the magnetization spinning in the transverse plane (and hence the emitted signal) diminishes. The rate at which this happens in various tissues can be demonstrated by "spin echo" pulse sequence, in which a 90° pulse is

followed rapidly by a 180° pulse. This pair of pulses is then followed by a brief burst of signals known as a "spin echo", from which an image can be made. If the time between the 90° pulse and the spin echo (which is known as TE) is relatively short (about 15-30 msec), the spin echo strength (and hence the image intensity) of various tissues will differ relatively little. But if the TE is long (60 msec or more), the strength of the spin echo emerging from different kinds of tissue will vary considerably, depending upon the tissue's T2 values, and will produce an image which is strongly T2-sensitive. Under these circumstances, tissues with long T2 values (which, like tissues with long T1 values, tend to be more "watery") will appear relatively bright, and vice versa.

All tissues of the urinary tract tend to appear to some degree darker than fat, which has a quite long T2. The different T2 values of normal and abnormal parts of the urinary tract differ from each other and enable the parts to be distinguished on T2-sensitive images. By these means, images of both normal anatomy and pathology can be created and so knowledge of the constituents of the tissues can be inferred.

In spin echo sequences, T1 weighted images are created by the faster moving protons and usually give better anatomical detail. T2 weighted images are delayed spin echo sequences and utilise the signals from slower moving protons. In this respect they may show

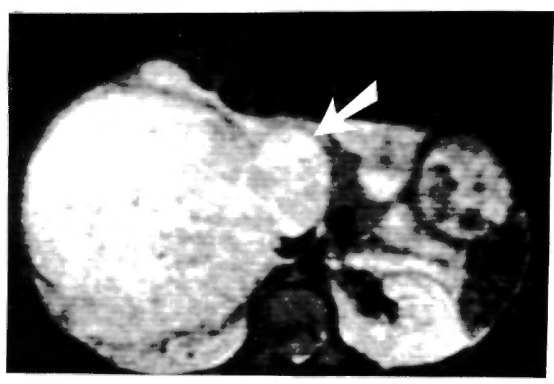
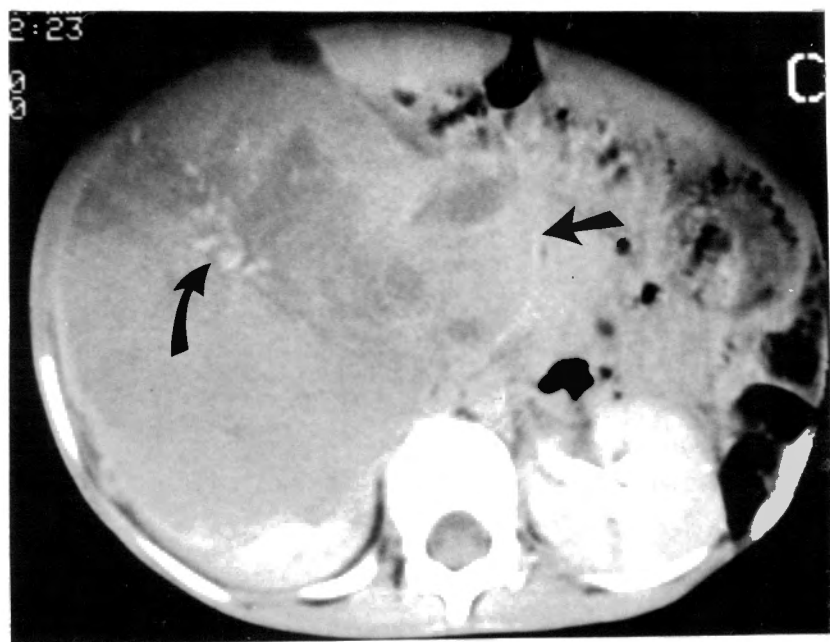
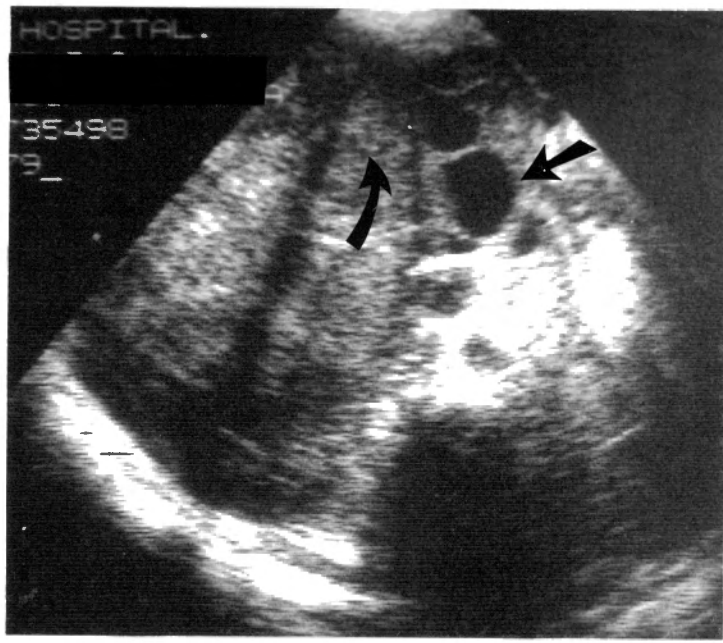
the variable nature and consistency of the tumour to better advantage.

Appearances

Our current experience is limited, but apart from being non-ionising MRI appears at this stage to have few advantages over CT and in many cases the image is inferior. We have illustrations of only two cases, both of which are shown in Fig. 16. The first case illustrates a comparison between US, CT and MRI, and all the images are taken roughly in the same transverse plane. All the information that was eventually obtained was available on the initial US scan. This showed that the mass had a medial exophytic bump which had cystic components. CT and MRI confirmed this, the latter showed that the "fluid" components in the bump give higher intensity signals in the T2 weighted images. There was no evidence of capsule invasion and none was found at surgery.

In the second case, MRI was of value. The child had a massive abdominal tumour. This was initially considered to be enlarged liver, as the child was jaundiced. It was noted that there was a superficial cystic "sausage" palpable in the abdomen. This was not given any great significance either clinically or on US. The MRI showed the "sausage" to be the displaced gall bladder and that the porta hepatis was occupied by an enormous Wilms' tumour.

FIG. 16



Case 2. Comparison of US and MRI in same child. The boy was referred to the hospital with jaundice and a large right abdominal mass. The presumptive diagnosis was a hepatic tumour. It was noted that there was a cystic sausage-shaped structure palpable in abdomen. The significance of this was not noted at the time and it was thought to be a possible loop of compressed bowel.

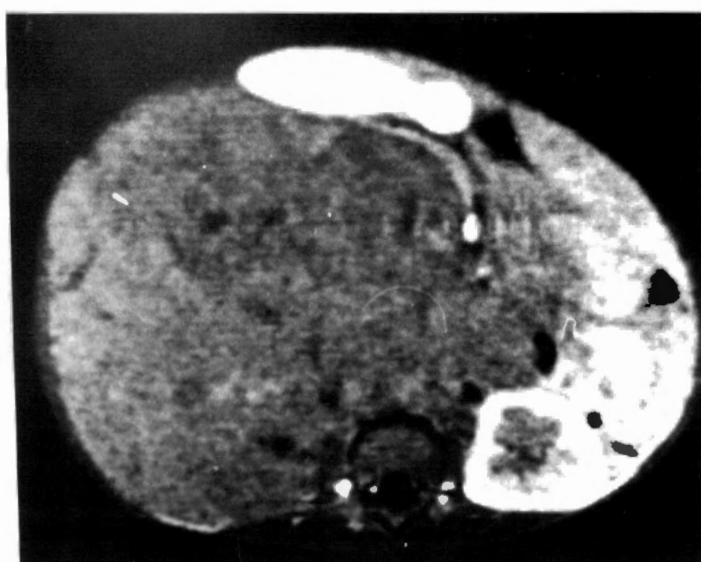
(a) Transverse US showing large right renal mass.

(b) Transverse US showing that there is a cystic structure anteriorly (arrow) and that there are also dilated cystic structures at the porta hepatis.

(c) MRI T2 weighted image (TR1700 TE66) shows dilated portal veins in the liver and shows that the previously noted cystic structure is gall bladder and that the cystic duct is grossly compressed.

An unusual case of Wilms' tumour causing porta hepatis obstruction and jaundice - confirmed as currently irresectable at surgery.

FIG. 16 continued



ARTERIOGRAPHY
&
NUCLEAR MEDICINE

Arteriography

The two modalities of arteriography and nuclear medicine will be considered together in this section. The reason for this is that both have a limited and similar use. This is to provide the surgeon with a functional image when it is required to conserve the normal parts of the affected kidney.

Our experience occurred during the period 1967-1971 when routine preoperative radiation to the tumours was employed. The purpose of the examination was to ensure a more exact diagnosis when the IVU findings offered any doubts. We reported 13 cases (Cremin & Kaschula) which at the time was the largest reported series. The other major report was by McDonald & Hiller from Royal Children's Hospital, Melbourne in which the author was also initially involved.

Technique

The examination is performed using a similar basal sedation as that outlined for CT. A routine Seldinger approach with local anaesthetic in the femoral region is employed. The technique used was described in the appendix of the publication by Cremin & Kaschula.

All the arteriograms were done by the femoral route under local anaesthesia and heavy basal sedation. There were no complications. The basal sedation was given intramuscularly, one hour before commencement and consisted of vallergran, 1mg per lb., droperidol 0.1mg per lb and morphine 0.1mg per lb. This sedation has a

prolonged effect and is effective provided there is no painful stimulus. It is necessary to restrain the child's legs for a short time at the end of the free aortic injection. The majority of the cases were aged between two and three years so that our catheters were standardized to a 19 gauge, thinwall Cook needle and 0.025in guide wire. The insertion of the catheters was subcutaneous except in two cases when a cut down procedure was performed so that the artery could be identified before direct puncture.

A free aortic injection through a French 5 Teflon catheter, external diameter 0.065in. (1.67mm) internal diameter, 0.039in. (0.99mm), was given. The injection consisted of 20ml of warmed Conray 280 given by a pressure pump that delivered the bolus over approximately two seconds.

Selective arteriography using 4ml of Conray 280 by hand injection was attempted in all cases and this only failed twice, when the renal arteries were grossly displaced and obturated. Usually selective arteriography was not difficult and a thin walled polyethelene catheter was used. (Cooks or Becton and Dickinson's polyethelene catheters, of internal diameter 0.056in. (1.67mm), external diameter 0.045in. (1.14mm), were satisfactory)."

Arteriography is now uncommonly performed but we would use basically the same technique apart from using

nonionic or low osmolality contrast media such as Hexabrix. These can be used in half the dosage and do not cause unpleasant side effects such as the hot sensations in the legs. Arteriography can be useful as a preoperative requirement in cases of solitary kidney (Fig.17) or bilateral tumours. The blood supply to normal and abnormal areas will be demonstrated. Tortuous tumour vessels of a spider leg nature may be seen and in large tumours there may be collateral circulation from adjacent lumbar vessels. The extent of the neovascularity will help in the preoperative assessment of the extent of the tumour.

Nuclear Medicine

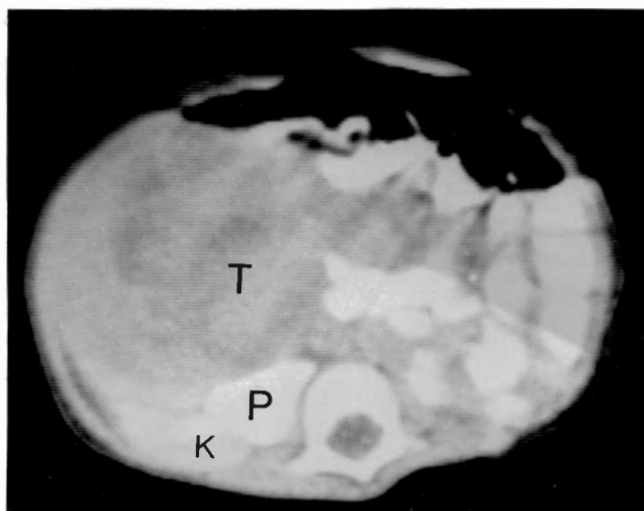
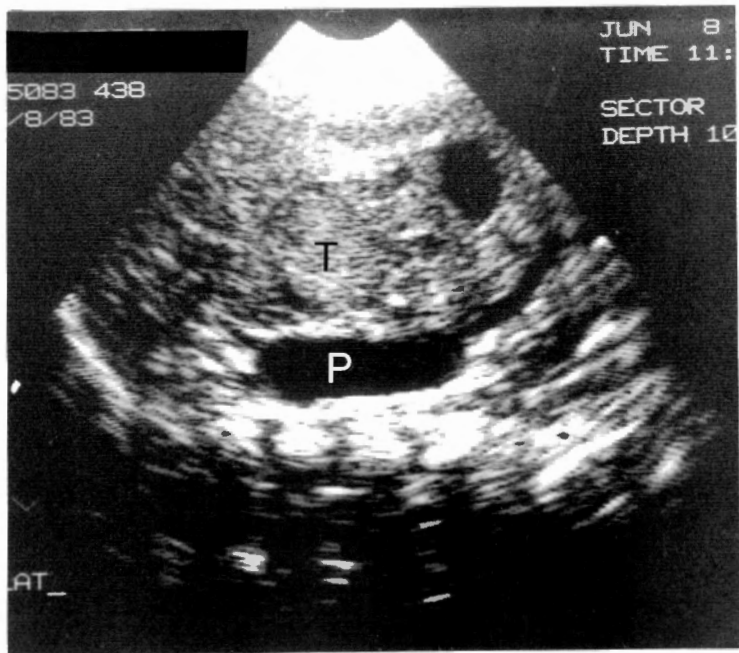
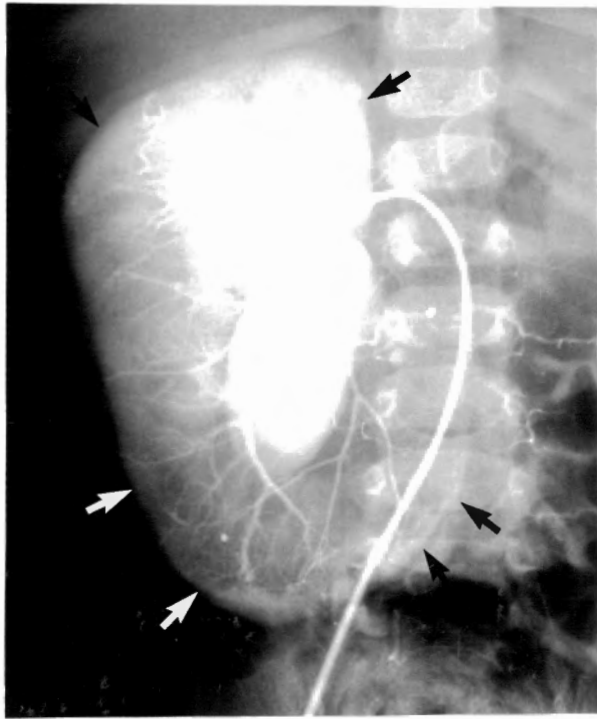
The isotopes used at our hospital for visualising the kidney are usually Tc99 D.T.P.A. which gives an image reflecting the glomerular function and the Tc DMSA which gives a delayed image reflecting tubular function. The indications for the use of nuclear medicine are roughly the same as those for arteriography so that the investigation may be performed in cases of solitary kidney or in synchronous bilateral tumours. In this respect a ^{99m}Tc D.M.S.A. scan will demonstrate the volume of functioning renal tissue and act as a guide to what may be preserved when bilateral surgery is contemplated (Fig.18). Scintigraphic evaluation using ^{99m}Tc glucoheptonate may also be used to distinguish true renal masses from normal variants (Sty, Starshak et al).

(a) This patient had a single right kidney and an arteriogram was performed to show the blood supply and the extent of the lesion (arrow).

(b) US demonstrates anterior circumscribed mass causing compression of the pelvis.

(c) CT scan confirms these findings. (T) tumour, (P) renal pelvis, (K) kidney.

FIG. 17





$^{99m}\text{TcDTPA}$ scan of bilateral Wilms' tumours. There are cold areas in both kidneys. The left kidney contributes 42% of overall renal function.

FOLLOW UP

Follow Up

The suggested protocol is designed both to investigate metastasis and to check "potential" cases.

1. Investigation of Metastasis

Metastatic secondaries usually spread to lung, bones and liver, though haematogenous spread may involve other parts of the body. The commonest site for secondaries is the lungs.

Metastatic lung lesions are rounded and may be single or multiple. Circumscribed deposits may be successfully removed by surgery so that the early detection of pulmonary metastases is important and critical in tumour management. They are often peripheral or subpleural in position (Fig. 19) which may make them difficult to detect by routine chest radiography.

In the past the lungs have been investigated by multi-positional chest radiographs and whole lung tomography. Recently it has been advocated that the initial chest radiograph and subsequent follow up should be replaced by CT (Brasch et al) and that the preoperative CT is essential for the proper staging of a Wilms' tumour (Sty et al).

Our facilities for chest C.T. are limited by necessity, so that our procedure for chest follow up is as follows (see also Appendix III).

Chest

We obtain a base line chest CT as soon as possible.

FIG. 19



CT scan of typical subpleural metastasis (arrow).

If CT is not employed for the initial diagnosis, then a CT of the chest is obtained as soon as is convenient and definitely before the end of the first period of chemotherapy. Our follow up protocol is routine chest radiographs at six weeks, three months and every three months for the first 24 months, followed by every six months for three years and yearly until puberty. If at any time the chest radiograph shows a suspicious lesion, the lungs are again checked by CT examination.

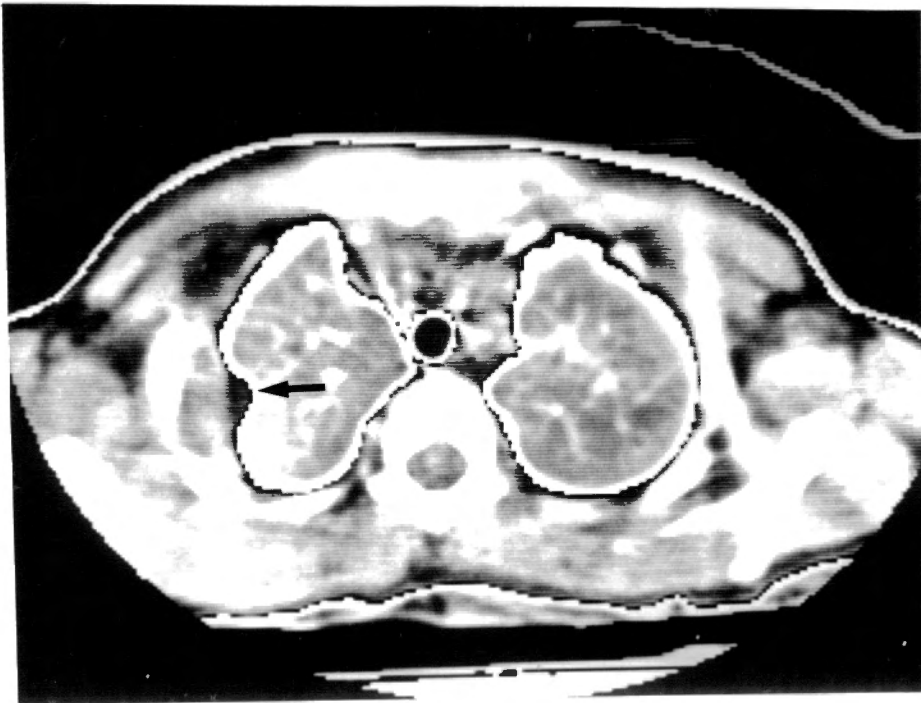
Unusual chest complications such as cavitation of secondaries and pneumothorax have been reported (Siegel & McAlister). Pleural effusion may occur and massive involvement may opacify a whole hemithorax.

Bones

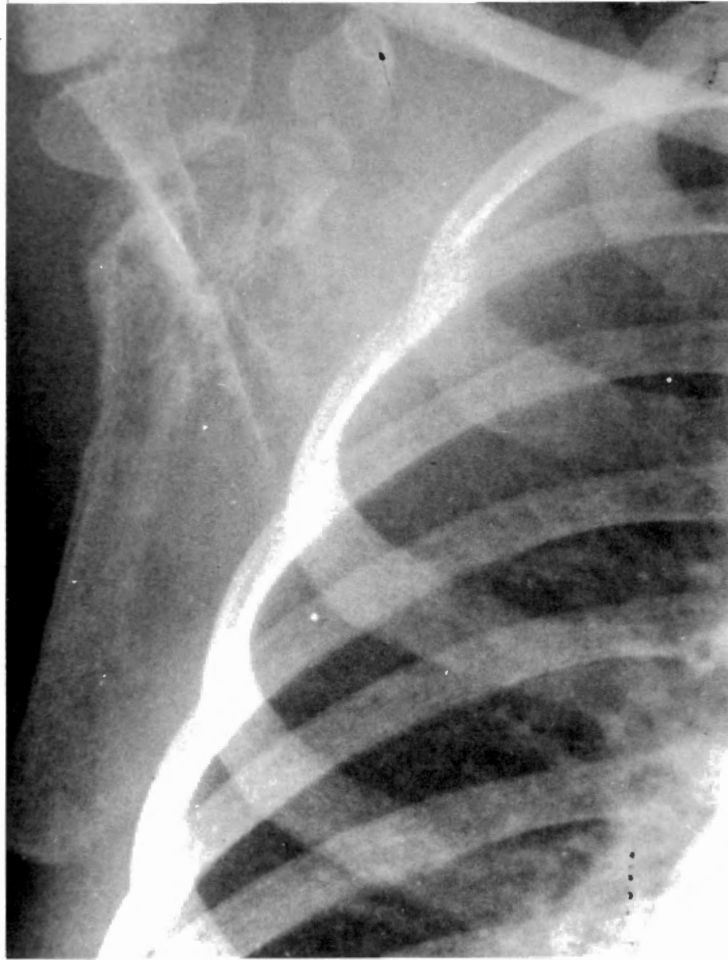
Metastases to bones are usually associated with the anaplastic or sarcomatous type of tumour (Marsden et al). It would be logical to restrict an initial complete skeletal survey to those who have this type of tumour. However our present policy is to perform an immediate skeletal survey on all cases. It is uncommon for there to be bone metastasis at the initial presentation but pathologists may wish to review their histology if bony lesions are detected (Lamego & Zerbini). The bone lesions are either diffusely lytic (Fig. 20) or more discrete circumscribed lesions. Both appearances may be seen in the same patient (Fig. 21). RNS is also used to evaluate the skeleton and may show



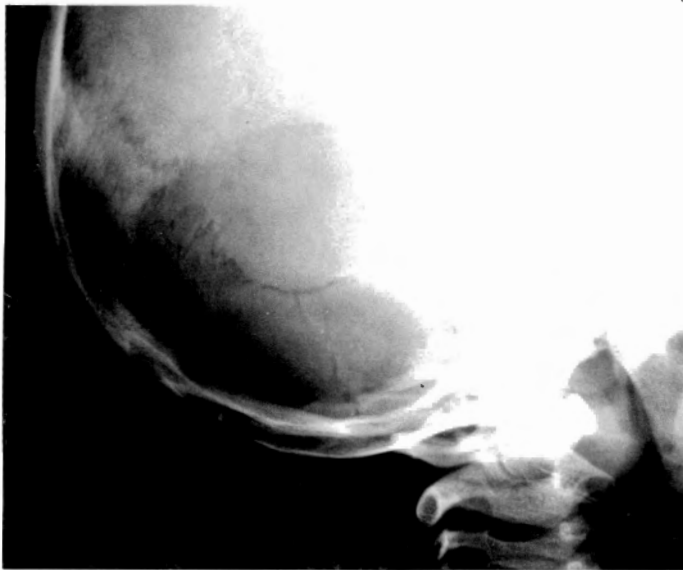
(a) Tomograms showing metastatic lytic lesions in the second right rib.



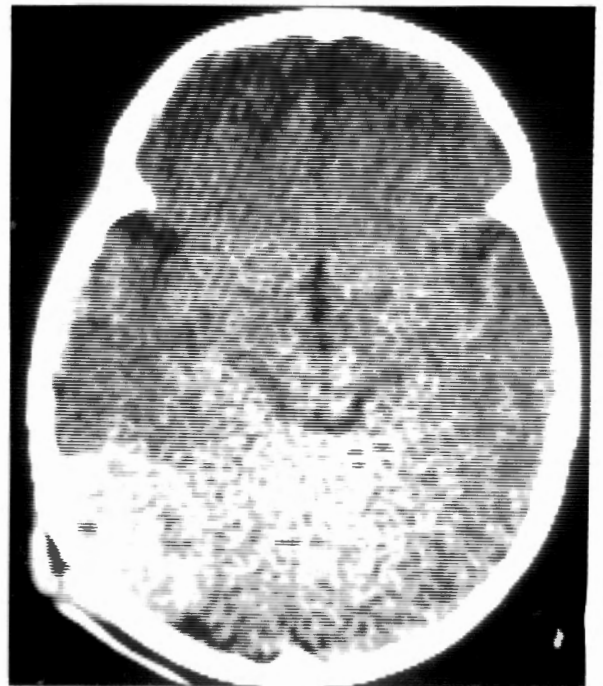
(b) CT scan of the same patient showing extra pleural invasion of the right thorax (arrows).



(a) Moth eaten lytic metastasis to right scapula.



(b) Same patient, lytic circumscribed lesion in bony occiput.



(c) CT showing bone destruction and extradural extension of tumour.

either photopenic areas or areas of increased uptake when there is abnormal osteoblastic activity.

Other areas

Both CT and US (Filiatrault et al) may be used to investigate recurrence of glands, complications from local spread and liver metastases (Fig. 22). Liver metastases are considered the second most frequent secondaries (Sty, Hernandez et al) but our experience has mainly concerned terminal cases. A large right sided kidney tumour may make an external impression on the liver that may simulate a metastasis on CT (Fig. 23). The use of ^{99m}Tc -Sulphur-colloid scintiscans may help resolve equivocal CT or US findings but they are also subject to the same problems of differentiating intra- from extra-hepatic lesions. MRI does not appear to be more effective than CT in visualising metastasis. Hepatic secondaries may be almost isointense on T1 weighted spin echo pulse sequences so that T2 weighted sequences or inversion recovery studies must also be utilised (Belt et al).

Kidneys

Tumours which subsequently develop in the contralateral kidney are more likely to be separate or metachronous primaries rather than the result of secondary spread and we perform a follow up IVU or US of the contralateral kidney for a further three years.

2. Complications

The follow up includes noting any adverse sequelae

FIG. 22

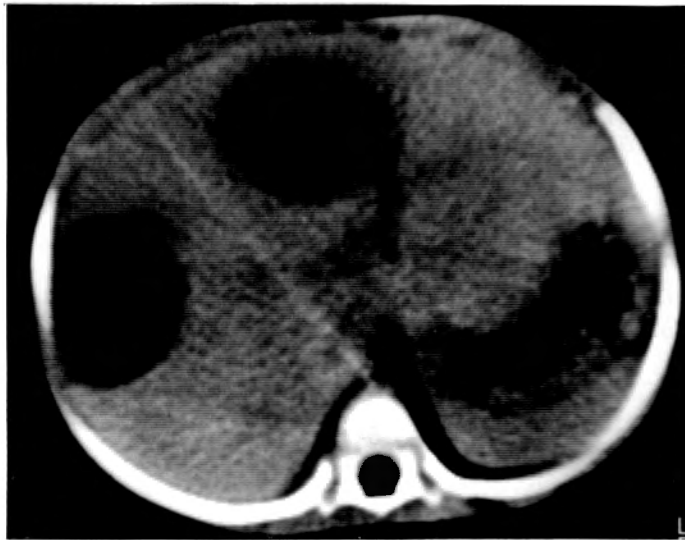


- (a) CT follow up after removal of left Wilms' tumour. Bolus injection demonstrates aorta (A) and right kidney (K). Arrow shows recurrence of tumour lymphatic glands (arrow).



- (b) CT follow up after a right Wilms' tumour removal 18 months previously. The cursors are on ascitic fluid which is displacing the liver. Arrow demonstrates a liver metastasis. Aorta (A) and left kidney (K).

FIG. 23



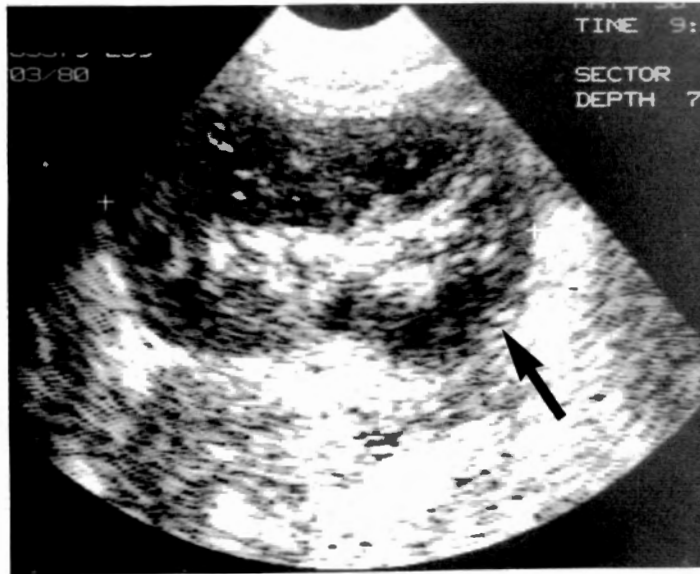
CT scan showing low density filling defects in liver due to external compression by large right Wilms' tumour (same case as in Fig. 2).

of treatment. The complications of treatment are mainly due to radiotherapy and the potentiating effects of chemotherapy (Lanzkowsky). Radiotherapy is currently usually reserved for patients that are at stage II and beyond. Keloid lesions in the bowel causing postoperative obstructions may occur as a complication of management. We have seen this only once. The orthopaedic sequelae that may result include avascular necrosis, severe scoliosis and limb-shortening (Katzman et al). Careful collimation has made these complications less frequent but aggressive therapy may cause damage to liver and the other kidney. Liver and renal function tests are therefore performed every six months for a two yearly period.

3. At risk cases

The investigation of potential cases with conditions such as sporadic aniridia or hemihypertrophy needs careful evaluation and follow up. It has been suggested that US is preferable to IVP as it is more likely to show early in the subcapsular cortex (Friedman). Our present policy is to assess these by US of the kidneys every six months up to the age of seven years. Even this is imperfect and errors may occur when the initial lesion does not cause enough disturbance to be detectable (Fig. 24). There is a case for more frequent US and possibly this should be performed every three months during the dangerous age period.

FIG. 24



- (a) Posterior sagittal US of right kidney in patient with hemihypertrophy. Multiple scans were performed which, even in retrospect, were considered normal. Tumour arose from site marked with arrow.



- (b) Anterior sagittal US seven months later showing the development of an anterior Wilms' tumour (T) from lower pole of kidney (K).

PROGNOSIS & SUMMARY

Prognosis

The prognosis for this remarkable tumour has changed to a 80% survival compared to a 80% mortality sixty years ago.

An account of the progress of treatment has been given by Johnson in his Chapter of Wilms' tumour in the textbook by Holden and Ashworth. It has more recently been highlighted by Kaufman (1986). His account notes that the advances in treatment is one of the major success stories in pediatric surgical oncology. In the early 1900s when nephrectomy alone was the treatment, the survival rate was about 15%. By the 1930s the improvements in anaesthesia and intensive care enhanced the rate to about 30%. By 1940 postoperative radiation had improved results to about 50%. In the mid fifties, Actinomycin D was first used and this, with external beam radiation has produced cures in the range of 80%.

The prognosis depends on the pathological pattern, the age at time of diagnosis and the extent of the disease. CT and MRI in the future may predict the extent of the disease far better than conventional radiology, but the final evaluation must be done at surgery and will depend on histology of surrounding tissues. It is this that will modify the staging and treatment of the condition.

OTHER TUMOURS

Mesoblastic Nephroma

This tumour may be suspected when an intrarenal mass is encountered in the first four months of life. It has also been labelled as a hamartoma that consists of a mass of dense fibromuscular bundles with no capsule that appears to give a locally infiltrative appearance. In general they are solid tumours but cystic regions may occur and some areas may show a high mitotic index (Snyder et al). There is nothing specific about the IVU (Fig. 25) and the U.S. shows a mainly uniform hyperechogenicity (Hartman et al).

Adenocarcinoma

This is a very rare tumour in the first decade of life but may be suspected when an intrarenal mass occurs in children beyond the age of 7 years. During the period under consideration there were only 2 cases at the Red Cross Children's Hospital (Kaschula). Pathologically the tumour is similar to that which occurs in adults (Hartman et al). Children commonly present with haematuria, abdominal pain or abdominal mass (Dehner et al) and this classical triad may occur in 15% of cases. The appearances on IVU will be the same as in Wilms' tumour though calcification is considered to be slightly more common. US and CT will also show an intrarenal mass and differentiation of renal cell carcinoma from Wilms' tumour ultimately depends on histological examination.

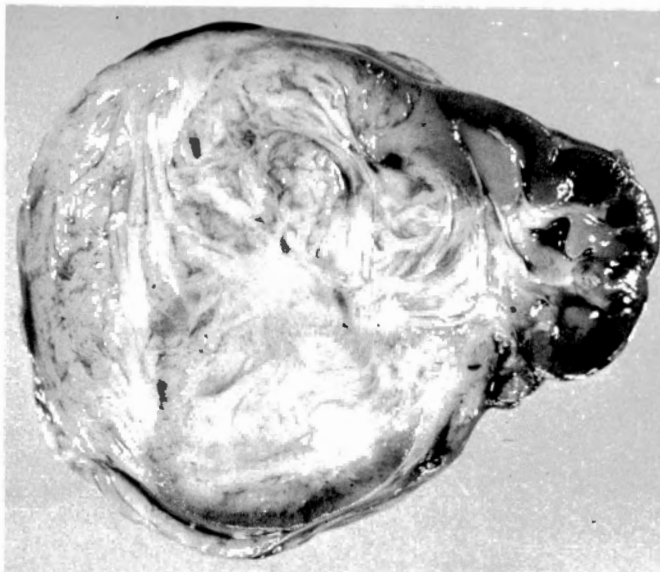
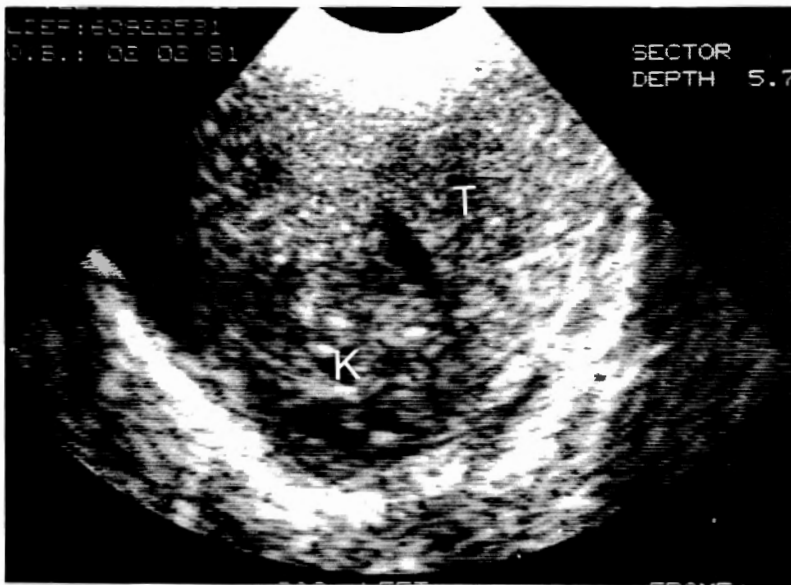
Chan et al reviewed the appearance of renal cell

(a) IIV tomogram in a five month old child with a mass due to mesoblastic nephroma.

(b) US showing the homogeneous tumour (T) arising from lower pole of kidney (K).

(c) Postoperative specimen showing the whorled fibromuscular bundles.

FIG. 25



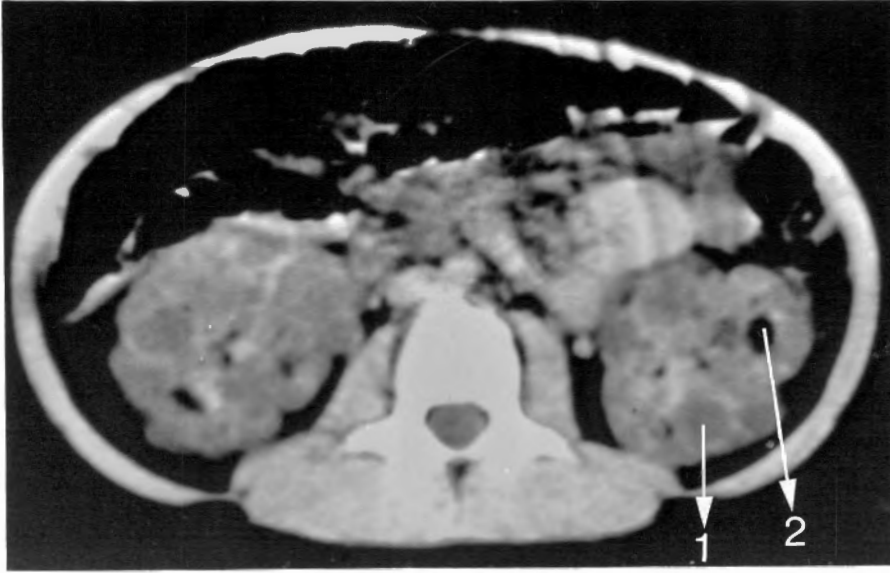
carcinoma in 17 cases that presented in the first two decades of life at the Toronto Children's Hospital. The mean age was 12 years. All the patients presented with signs and symptoms referable to the primary tumour. In comparison to Wilms' tumour, the renal cell carcinoma in childhood and adolescence usually presented with a much smaller abdominal mass which is frequently not palpable or visible on plain abdominal radiographs. The US and CT showed non specific solid intrarenal masses.

Miscellaneous

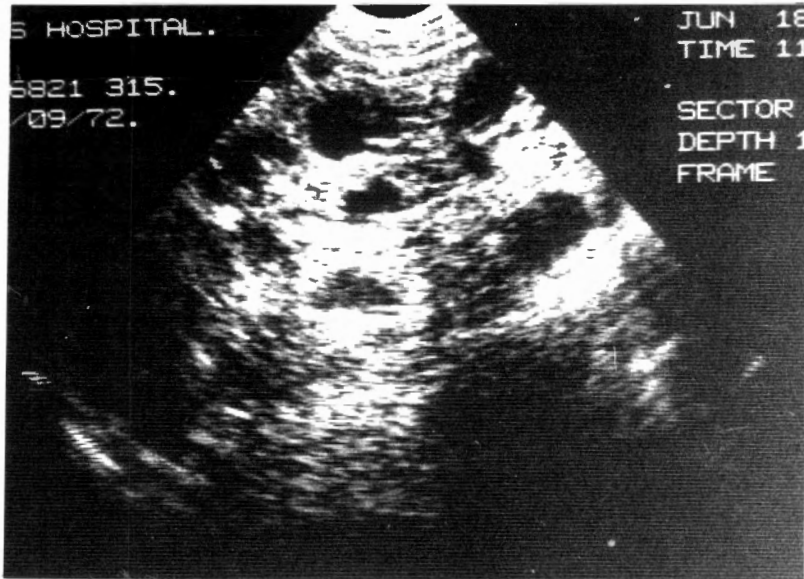
Angiomyolipomas (Sherman et al), rhabdomyosarcomas (Lifschultz et al) and haemangiomas (Samellas et al) are all rare tumours in childhood. Angiomyolipomas are usually associated with tuberous sclerosis in which skin and skeletal lesions are combined with mental retardation and intraventricular calcification. The tumours are bilateral and on US the fatty elements will cause a highly echogenic lesion. On CT the fatty density of the tumour will cause areas of below zero attenuation.

It should be noted that the detection of fatty elements by CT does not necessarily imply a benign tumour as fat may also occur in Wilms' tumour (Parvey et al). The tumours may be cystic rather than solid (Mitnick et al) and may cause renal failure (Fig.26). The tumours are classified as "benign" but may be locally aggressive, replacing kidney tissue and extending

FIG. 26



- (a) CT of a 5 year old boy with tuberous sclerosis. Both cystic (1) and fatty low attenuation (2) elements are present.



- (b) Sagittal US of kidney showing the cystic nature of the mass.

into prevertebral tissues and local lymph glands. They may also be associated with the later development of a renal cell carcinoma (Gutteriez et al).

SECONDARY TUMOURS

Secondary Tumours

The kidneys are quite commonly involved in disseminated lymphoma or leukaemia. In lymphoma the infiltration may cause either diffuse renal enlargement (Fig. 27) or nodular distortion of the calyces. The nodular nature of lymphoma may be demonstrated on US as discrete hypoechoic areas or on CT as focal areas of reduced attenuation. Multiple glands around the aorta will show as round, compact hypoechoic areas and on CT they are best demonstrated by a bolus technique. In this form of dynamic study the aortic vessels are enhanced by an injection of contrast medium immediately prior to and during the scanning. Leukaemic involvement of the kidney does not usually produce nodular lesions but causes bilateral enlargement with uniform compression of the calyces on US. It gives a non-specific generalised loss of echogenicity with loss of corticomedullary margins. In both lymphoma and leukaemic infiltration, RNS using ^{67}Ga may be helpful (Shirkhoda et al).

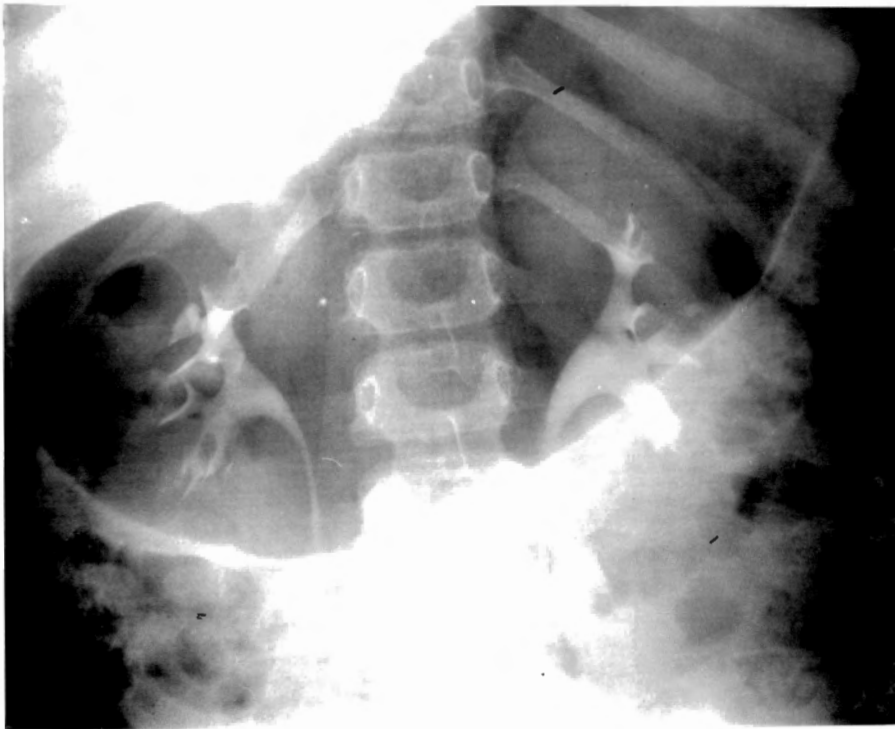
Tumours of Renal Pelvis and Ureter

These are all rare and consist mainly of pedunculated fibro epithelial polyps (Colgan et al). When suspected their nature and mobility may be demonstrated by retrograde urography. From Bern in Switzerland an account of three cases is given by Tschappler and Fishedick. They were investigated by IVU and showed smooth oblong filling defects in the

FIG. 27



(a) IWU showing bilateral displacement of the calyceal systems due to diffuse infiltration by lymphoma.



(b) Normal appearance after three months chemotherapy.

proximal ureter, with and without finger like extensions and were associated with mild pyelocaliectasis. No malignant potential existed in the tumours. Other rare lesions that have been reported are leiomyoma of the ureter (Mondschein et al), cholesteatoma (Ross et al) and haemangioma of the renal pelvis (Vinour et al).

Extrarenal Wilms' Tumours

For the sake of completeness this entity will now be considered.

An extrarenal Wilms' tumour is a most uncommon condition. The term may refer to a tumour apart from, but adjacent to the kidney. Its more correct use is for the rare tumour that has nephroblastoma histology found in the mediastinum, retroperitoneum or along the path of testicular descent (Orlowski et al; Madenat et al). We have had no experience of this lesion during the last twenty years at the Red Cross Children's Hospital. On the one occasion when we considered it a possible diagnosis in a para renal mass, the pathologist revised his opinion to that of an atypical neuroblastoma.

LOWER TRACT TUMOURS

GENERAL SURVEY OF LITERATURE AND COMMENTS

Tumours of the bladder in children are rare, so that there is no great volume of publications on imaging. Going back into the literature, one of the earliest surveys of their occurrence was in 1924. This was by Deming who documented 65 cases in the first decade of life. These were classified as follows:-

Pathology -----	No. --	Per Cent -----
Sarcoma	37	59.9
Myxoma	16	24.5
Fibroma	5	7.6
Rhabdomyoma	2	3.0
Polyp	2	3.0
Dermoid	1	1.5
Papilloma	1	1.5
Myoma	1	1.5
	--	
	65	

If one makes allowances for the evolutionary changes that occur in pathological nomenclature the first four entities (92%) would currently be considered as part of the rhabdomyosarcoma spectrum.

In 1959 Thompson and Coppridge reported five cases of bladder sarcoma and added these to the literature of 80 reported cases. It was considered then that the key to survival was early diagnosis and aggressive surgery. Any case lasting as long as a year after definitive treatment had a good prognosis.

In 1970 the radiologists from the University Hospital of Cleveland and Case Western Reserve University, Ohio, reviewed their experiences with cystography in bladder tumours of childhood (Tucker &

Persky). I have been in contact with Dr. Tucker as his report is one of the few in the radiological literature. In 16 years they had encountered 11 bladder tumours in children below the age of 16 years. One of these cases was originally considered histologically benign but by its subsequent behaviour was found to be malignant. The cases are shown in Table V.

Their method of imaging was cystography and all the tumours presented as filling defects. Benign tumours may have an extensive intramural growth so that they may be indistinguishable from malignant growths. The differential diagnosis of these filling defects included foreign bodies and ureterocoeles.

Our experience of cystography has been similar but I would add to the differential diagnosis, blood clot in severe cystitis and granulomas in chronic infections such as tuberculosis and bilharzia.

Any intramural extension with bladder displacement usually indicates malignancy but it may not be possible to distinguish a bladder tumour from other expanding pelvic tumours. A large sacrococcygeal teratoma may lift the bladder out of the pelvis in a similar fashion to a prostatic rhabdosarcoma. Other conditions that may cause bladder displacement on a cystogram would be pelvic abscesses, ovarian cysts and hydrometrocolpus.

The following observations on this literature are pertinent. Up till 10 years ago the cystogram was the

TABLE V

No.	Sex	Age	Diagnosis
Benign			
1	M	2y 5m	Angiohamartomatous malformation
2	M	7y	Hemangioma
3	F	10y	Transitional cell papilloma
Malignant			
4	M	4y	Sarcoma
5	M	1m	Sarcoma
6	M	1ly	Originally, epithelial polyp; eventually sarcoma
7	F	1y 9m	Sarcoma
8	M	3y 6m	Sarcoma
9	M	7y	Sarcoma
10	M	4y	Mesenchymoma
11	F	4y	Sarcoma

There were two survivors in the malignant group.

Adapted from Tucker & Persky.

main method of diagnostic imaging of pelvic tumours. Since then cystography has been relegated to a more secondary place as it has been largely replaced by CT and US. It is difficult to make any comparison between current methods of imaging and those that existed previously as there is no volume of literature on the subject.

A comparison of different methods of imaging of paediatric abdominal masses by Kirks et al is a general overview of the subject with only limited reference to the use of CT in the pelvis. In their communication about MRI, Cohen et al refer to one case of pelvic rhabdomyosarcoma, noting the value of sagittal scans in assessing the response to chemotherapy. Although we only see these tumours about once every two years, the modern imaging techniques have shown themselves superior to the traditional cystogram. A brief examination by ultrasound can now ascertain whether the suspected pathology is present and if it is either tumour or non tumour. After that a decision about the use of other techniques to best outline the tumour and its response to treatment can be made.

Rhabdomyosarcoma

The commonest soft tissue sarcoma in childhood is the rhabdomyosarcoma, of which about 20% occur in the urogenital tract (Exelby et al). They are usually embryonal or alveolar in nature and have a botryoid sub-group. The botryoid tumours are formed when

proliferation occurs into a hollow cavity to give a polypoid grape-like appearance. There is no common agreement as to the tissue of origin of these tumours (Ghazali), but they are twice as common in boys than girls. In a series of 20 rhabdomyosarcomas collected by Williams & Schistad there were 14 from bladder and 6 from the prostate, 9 of the bladder tumours survived after radical surgery, whereas none of the prostate tumours survived. In the pelvis they usually present before the age of 3 years and involvement of the bladder base gives dysuria, retention and urinary tract infection. They are locally extremely invasive with later metastasis via lymphatic or haematogenous routes. The treatment is by anterior extirpation, radiotherapy and combined chemotherapy. The Intergroup Rhabdomyosarcoma Staging (IRS) at time of diagnosis is given in Table VI (Maurer et al).

Our experience of this tumour has been extended to cover the 10 year period 1976-1985. We have not examined our cases before that period as we did not have use of US facilities until 1976. During this period there were 5 cases of rhabdomyosarcoma of which 3 cases are still surviving (Karabus).

TABLE VI
-----STAGING

At the time of diagnosis, the patients were staged according to the Intergroup Rhabdomyosarcoma Staging (IRS) classification:

Stage I - Completely resected localized disease.

Stage II - Grossly resected tumour with regional disease in the form of positive lymph nodes and /or microscopic residual disease.

Stage III - Gross residual disease.

Stage IV - Metastatic disease.

Reference: Maurer et al

IMAGING MODALITY

Imaging Modalities

Cystography

Radiographs of the contrast-filled bladder may be obtained either as part of the IVU series or by direct instillation by catheter during a micturating cystourethrogram. Both may show intravesical filling defects or displacement of the bladder. The IVU series has the advantage of also showing displacement or involvement of ureter and obstruction. Delayed films (1 to 2 hours) to show the kidney involvement should be taken when obstruction exists. The examination may be combined with a contrast examination of the rectum.

Ultrasound

US is an excellent method of imaging pelvic masses. It will also show either cystic or solid components in the tumour. It will show the relationship of the mass to the bladder, but an effective examination requires a reasonably full bladder. The bladder may have to be filled with water by a catheter as penetration of sound waves into the depths of the bony pelvis is not always possible if bowel distention is present.

US is an easy and non-invasive examination and it should be the first imaging procedure to be undertaken in cases of pelvic tumours. It may also be used in the follow up of cases, but in this respect CT and MRI may have imaging advantages.

Computed Tomography

It is in the extrarenal retroperitoneal and pelvic

application. It will demonstrate the extent and infiltration of the tumour and show its shrinkage and response to therapy. Careful technique includes bowel opacification by oral contrast media a few hours beforehand and rectal opacification by direct injection of water soluble contrast media immediately prior to the examination (Cremin & Mervis). Examples of the use of IVU, US and CT are demonstrated in Figures 28, 29 and 30, with the use of MRI in the follow up in Fig 30.

Magnetic Resonance

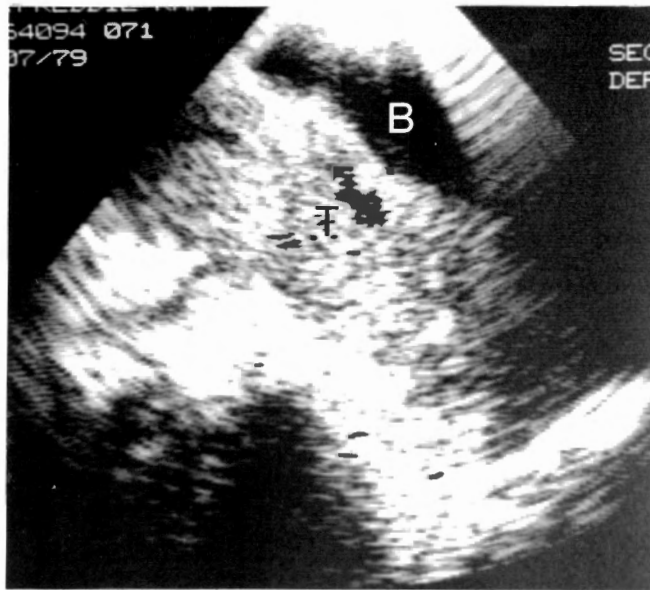
There is as yet no extensive publication on the use of MRI but it has great potential for defining the extent of soft tissue involvement. The properties of adjacent tissues with different structures such as bone, muscle, fat and urine in the bladder can be defined and separated by measuring their proton densities in both their T1 and T2 weighted spin echo properties. In this manner a more accurate knowledge of local invasion is obtained. Its use is shown in the follow up 2 years later of the case illustrated in Fig. 30. The ease by which multisections in coronal, sagittal and axial planes is shown in Fig. 31.

(a) IWJ in a four year old boy showing a large mass in the left pelvis that is displacing the bladder to the right and obstructing the left kidney.

(b) Sagittal US of the bladder (B) showing the invasion by solid tumour (T).

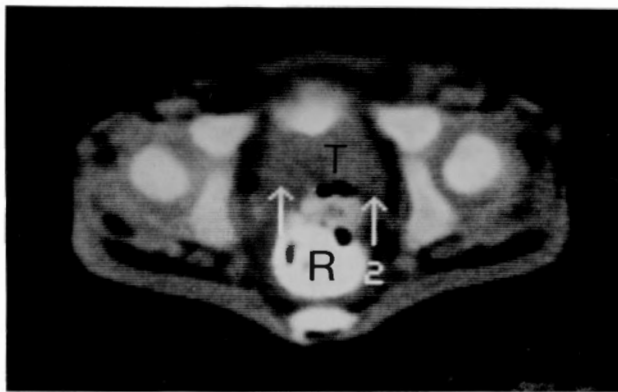
(c) CT six months after chemotherapy shows tumour (T) has become partially necrotic. Contrast medium is layering in the posterior aspects of the bladder.

FIG. 28





- (a) IWU of extensive rhabdomyosarcoma causing multiple "thumb print" filling defects in bladder. There is some displacement of lower ureters by glands.



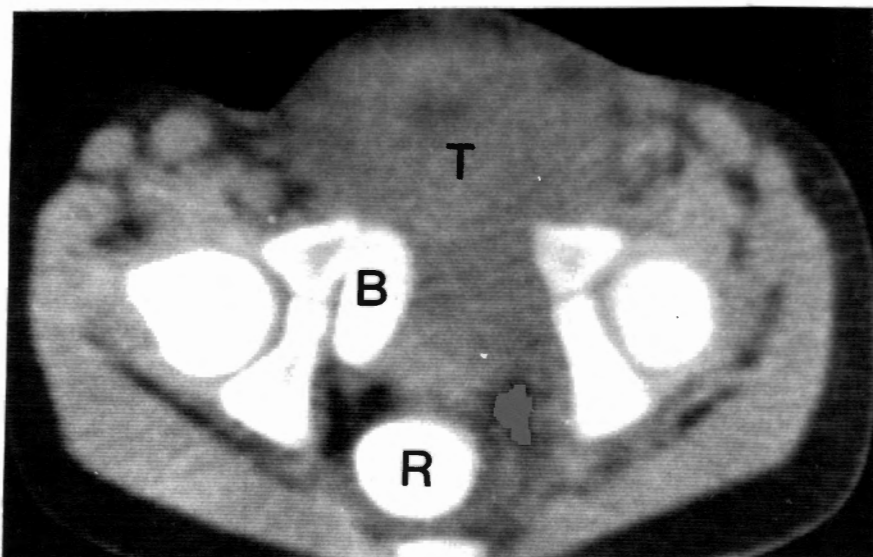
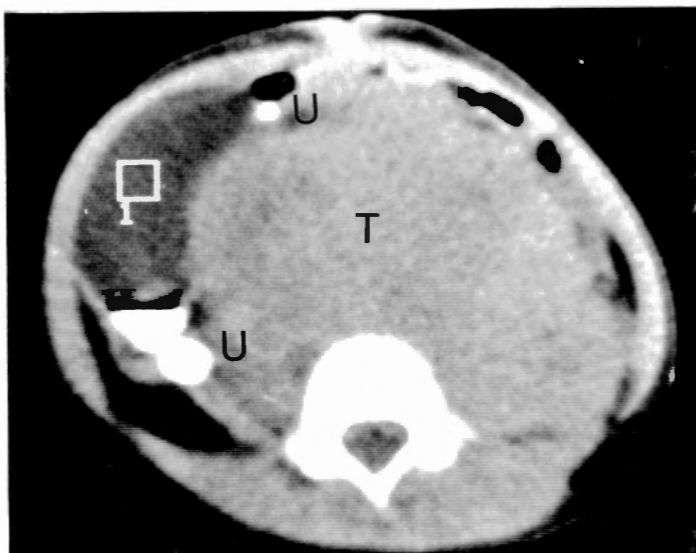
- (b) CT shows that the tumour (T) extends down between bladder neck and rectum (R). Contrast media was instilled into rectum to outline it.

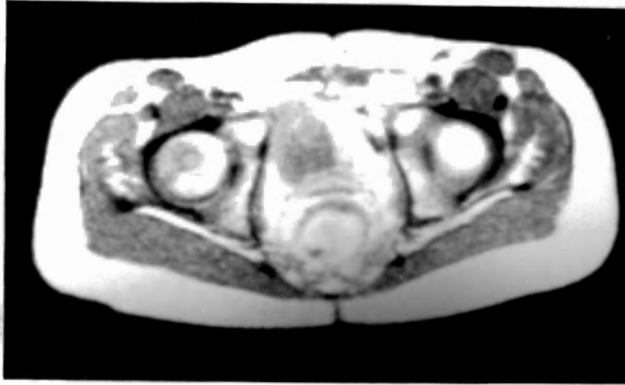
(a) A five year old girl with embryonal rhabdomyosarcoma of pelvis. The IWU (a) shows gross displacement of the bladder to the right and obstruction of the left kidney.

(b) CT scan showing the tumour (T) causing gross displacement of bladder (marked by cursor) and both ureters (U).

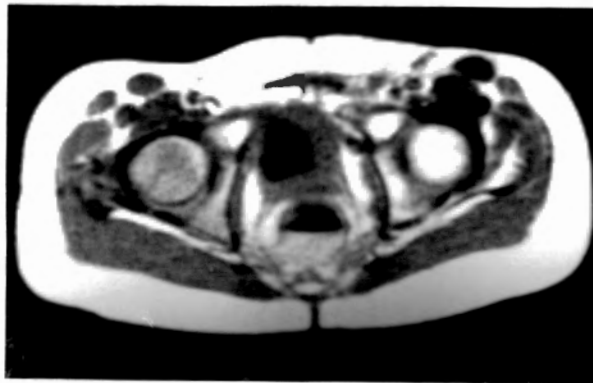
(c) CT scan through the lower pelvis shows that the mass is extending out into the abdominal muscles. Tumour (T), bladder (B), rectum (R).

FIG. 30





- (d) MRI 2 years later after irradiation and chemotherapy.
T1 weighted image (SE600/29).



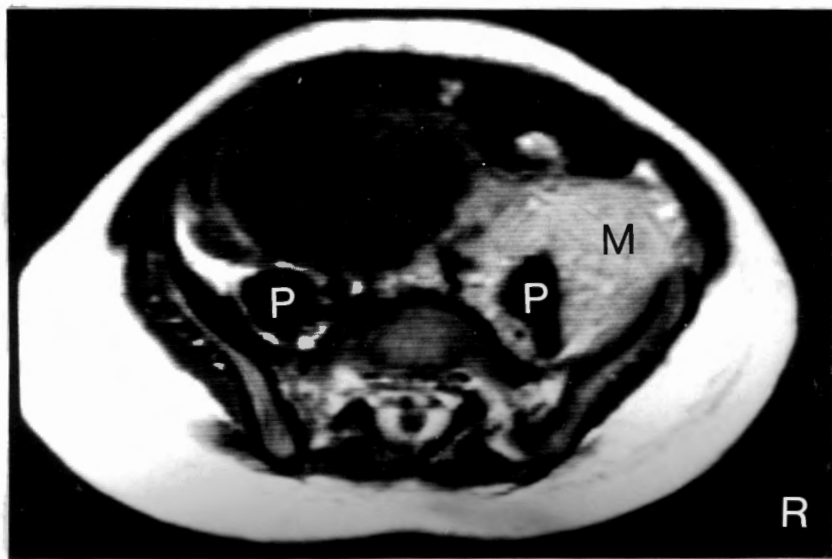
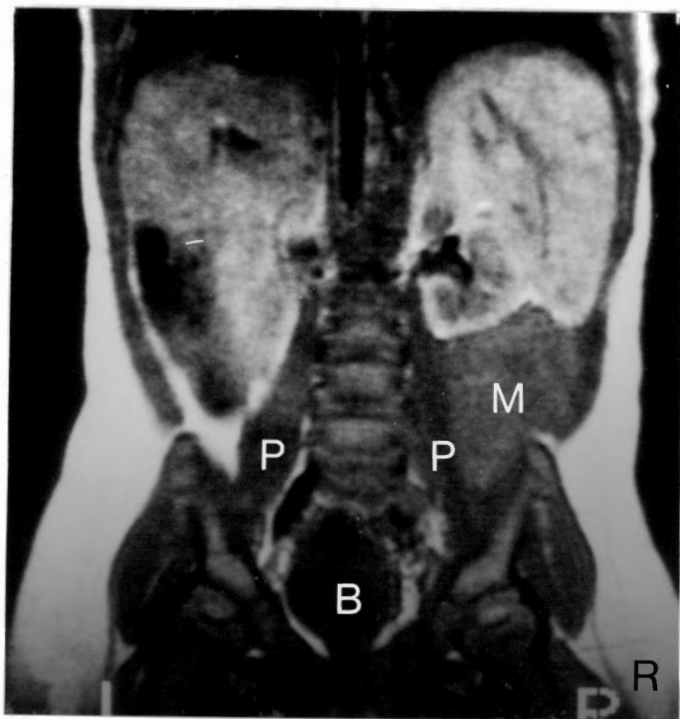
- (e) T2 weighted image (SE1700/29). Both these images show similar features. The bladder is slightly displaced to the right by some abnormal soft tissue. The soft tissue has the intensity on the T2 weighted image of being fatty. At surgery this was confirmed and no malignancy was found.

(a) MRI T2 weighted (TR1700 TE27) coronal image showing large mass (M) to the right of the psoas muscle (P) displacing kidney upwards and descending into the pelvis to involve right side of bladder (B).

(b) Sagittal section showing similar features and the upward displacement of kidney.

(c) Markedly weighted T2 (TR1700 TE90) image in transverse section of lower abdomen showing infiltration of mass around the psoas muscle (P). The acquisition of the slower T2 signals demonstrates tumour against the almost blacked out normal structures.

FIG. 31



Other malignant tumours

These are all rare. Low grade transitional cell tumours (Castellanos et al), leukaemic infiltration (Troup et al) and carcinoma of the prostate (Shimada et al) have all been reported. When there is bladder involvement the preoperative differentiation from a rhabdomyosarcoma without a biopsy will be difficult. Endodermal sinus tumours (yolk sac tumours) that arise from mesonephric duct remnants are very uncommon but may cause pelvic infiltration (Duckett et al). The "thumb printing" filling defects of the bladder that indicate mucosal infiltration may occur in non-malignant conditions such as severe cystitis. A haemorrhagic form of cystitis that causes marked mucosal thickening may result as a complication of cyclophosphamide administration. Granulomatous conditions such as tuberculosis and bilharzia that cause infiltration of the bladder should also be considered in endemic areas.

Benign tumour of bladder and urethra

Bladder benign tumours are rare and when Ganem & Ainsworth researched their incidence in 1955 they found only 30 cases. These were reported as follows:-

Hemangioma	9
Polyp	5
Neurofibroma	4
Fibroma	3
Fibromyoma	3
Other fibroma	3

We have seen only two benign bladder tumours in the last 18 years. These were fibroepithelial polyps that

arose from either the bladder base or from the posterior urethra at the verumontanum. They form a round filling defect at the bladder base during an IVU or cystogram. They are mobile so that those in the posterior urethra may float upwards into the bladder or they may be seen to prolapse downwards into the posterior urethra during micturition (Fig. 32). There has been one report of US diagnosis of urethral polyp (de Filippi et al). This was in a six year old boy who had a polyp with a long stalk that rose from the verumontanum. Its mobility into the bladder was successfully demonstrated.

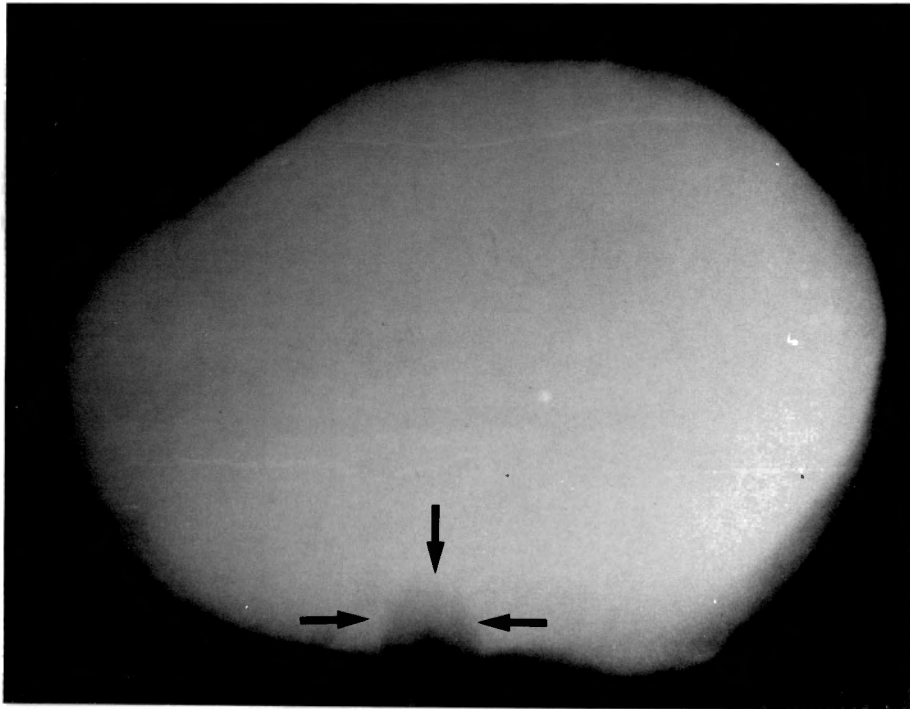
Neurofibromatosis

Although neurofibromatosis is a fairly common condition in Cape Town, we have no experience of involvement of the bladder in this condition.

Three children were reported by Daneman & Grattan-Smith from the Prince Alexander Hospital for Children in Sydney, Australia and during a visit some years ago I was shown one of the cases. They reviewed the literature and found 17 previously reported cases. The bladder was involved in males compared to females by a ratio of 3:1. Approximately one quarter had a family history of neurofibromatosis and two thirds had other manifestations such as cafe-au-lait spots (Daneman & Grattan-Smith).

The lesions may present few genito-urinary features apart from a pelvic mass or enlarged genitalia. Distension of the bladder may be due to obstruction by a

FIG. 32



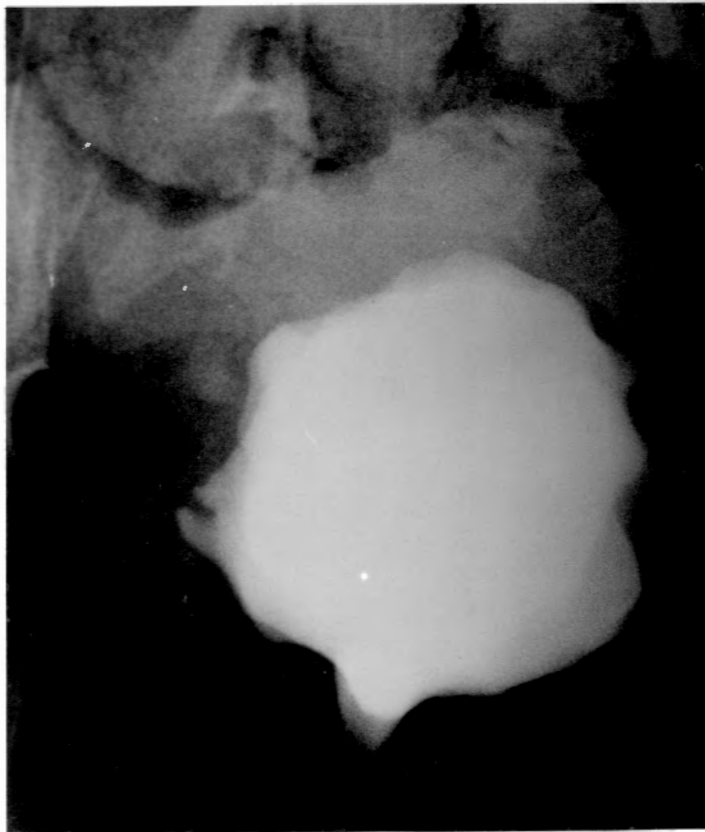
(a) and (b) Polyp (arrows) prolapsing down from bladder into posterior urethra.

tumour mass or by disturbance of the nerve supply.

Localised lesions may cause filling defects in the bladder which may be extrinsic or intrinsic (Figs. 33, 34 and 35). More diffuse lesions cause lobulation and irregularities of the bladder wall so that the bladder base and posterior urethra may be elevated.



Eight year old girl with stigmata of neurofibromatosis. Presented with recurrent abdominal pain and lower abdominal mass. The IVU shows large abdominal mass which is distorting the bladder and displacing the ureters.



A two and a half year old boy who presented with dribbling incontinence and a swelling of the penis. VCUG - post micturition film showing a large residual volume, irregular bladder wall, elevated bladder base and open bladder neck.

Acknowledgement to A. Daneman & Gratten Smith.
Pediatric Radiology 1976 4:166.

A two and a half year old boy with known neurofibromatosis who presented with abdominal distension.



- (a) IVU showed narrowed lower end of left ureter, irregular / lower end of right ureter and transradiant defects in the bladder.



- (b) VCUG showing filling defect at the base of the bladder.

Acknowledgement to A. Daneman & Gratten Smith.
Pediatric Radiology 1976 4:166.

CONCLUSIONS

Upper Tract Tumours

- 1) US has now become the primary diagnostic imaging modality for renal tumours.
- 2) US will demonstrate the intrarenal nature of the tumour. An intrarenal solid mass in the age group 1-5 years is a Wilms' tumour till proven otherwise.
- 3) US results are operator dependant and success depends on experience.
- 4) The IVU still has a definite place in the investigation. It will confirm the US findings and demonstrate the normality of the contralateral kidney.
- 5) Institutions should use the modalities that they are most practiced in. US and IVU are readily available so that they are currently the most widely used combination.
- 6) CT has not significantly altered our initial diagnostic capabilities.
- 7) CT is the best method for excluding lung metastases. It has advantages in the follow up of patients. We have been hampered in evaluating this by not having a unit on site. This will be rectified in 1987.
- 8) MRI has yet to demonstrate its superiority for routine use. Its orthogonal imaging capabilities are advantageous in the study of the inferior vena cava. In particular it is more helpful in distinguishing between caval displacement, invasion and obstruction by tumour thrombus.

Lower Tract Tumours

- 1) Ultrasound is the method of choice for primary diagnostic imaging.
- 2) There is still a place for the IVU as this will show the extent of bladder involvement and obstruction of ureter.
- 3) CT is the method of choice for the follow up of cases treated by drugs and radiotherapy.
- 4) MRI may have advantages in showing residual tumour but there is as yet no large series to prove this point.

Final Analysis

Having carefully researched our material and the literature on the subject, we return to the quotation in our dedication. There can be no truth without trial and it is self obvious that no imaging modality is without error, unless it is considered in its clinical perspective.

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APPENDIX 1RED CROSS CHILDREN'S HOSPITAL ONCOLOGY SERVICETREATMENT OF WILMS' TUMOUR - (Jan 1981)STAGE I

<u>SURGERY</u>	Day 1	Nephrectomy
<u>CHEMOTHERAPY</u>		<u>CHI</u>
Day 1		Actinomycin D 15mg/kg IV daily x 5
Week 1 - 10		Vincristine 1.5mg/m ² IV weekly x 10
Month 1 ,3,6		Actinomycin D 15ug/kg IV daily x 5
Month 3,6		Vincristine 1.5mg/m ² IV x 2 (on days 1 & 5 of AMD course)

STAGE II

<u>SURGERY</u>	Day 1	Nephrectomy
<u>RADIOTHERAPY</u>	Week 1	2000 rads maximum (reduce for age) to renal bed
<u>CHEMOTHERAPY</u>		<u>CHI</u>
Day 1		Actinomycin D 15ug/kg IV daily x 5
Week 1 - 10		Vincristine 1.5mg/m ² IV weekly x 10
Month 1 ,3,6,9,12,15		Actinomycin D 15ug/kg IV daily x 5
Month 3,6,9,12,15		Vincristine 1.5mg/m ² IV x 2 (on days 1 & 5 of AMD courds)

STAGE III

<u>SURGERY</u>	Day 1	Nephrectomy and accessible tumour resection.
<u>RADIOTHERAPY</u>	Week 1	2000 rads maximum to renal bed (reduce for age)
<u>CHEMOTHERAPY</u>		
	Day 1	Actinomycin D 15ug/kg IV daily x 5
	Week 1 - 10	Vincristine 1.5mg/m ² IV weekly x 10
	Month 1 ,4 ,7 ,10 ,13	Adriamycin 30mg/m ² IV daily x 2
	Month 3,6,9,12,15	Actinomycin D 15ug/kg IV daily x 5 Vincristine 1.5mg/kg IV x 2 (on days 1 & 5 of AMD course)

STAGE IV and UNFAVOURABLE HISTOLOGY (all stages)

<u>SURGERY</u>	as above
<u>RADIOTHERAPY</u>	to 3500 rads. Consider radiotherapy to metastases and/or surgical removal if accessible.
<u>CHEMOTHERAPY</u>	as above Add Cyclophosphamide 10mg/kg IV daily x 3 with ADR & AMD courses.

Acknowledgement: Dr. C. Karabus

APPENDIX 2

FOLLOW UP PROTOCOL

- 1) Physical examination every three months for 2 years, then every six months for 5 years.
- 2) Chest x-ray every six weeks for 3 months, then every 6 months for 3 years.
- 3) Chest CT at the beginning and the end of chemotherapy regime and as required as alternative to (2).
- 4) IVP or US every year for 3 years.
- 5) Liver and renal function every 6 months for 2 years.
- 6) Liver and bone scans as required.

When pulmonary metastases are present, chest x-ray repeated at 6 weekly intervals till normal for at least 9 months. Then every 3 months for another 2 years.

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APPENDIX 3SURVIVAL RATESWILMS TUMOURS 1968-1985

<u>Stage</u>		<u>Alive</u>	<u>1968-74</u>	<u>1974-80</u>	<u>1981-85</u>
I	55	41	11/3	13/14	16/18
II	18	12	4/6	3/7	5/5
III	25	17	6/9	5/8	6/8
IV	8	2	0/3	2/2	0/3
V	7	0			

TOTAL	113	72(63.7%)	21/41(51.2%)	23/31(74.2%)	28/34(82.4%)
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Figures given by Dr. C. Karabus