A Study of Lower Urinary Tract Anatomy

and

Selected Pathology

in

Boys

A Thesis for the Degree of Master of Surgery
in the University of Adelaide

PA Dewan
CERTIFICATION

This work contains no material which has been accepted for the award of any other degree or diploma in any university or other tertiary institution and, to the best of my knowledge and belief, contains no material previously written or published by another person, except where due reference has been made in the text.

I give consent to this copy of my thesis, when deposited in the University library, being available for loan and photocopying.

PA Dewan
MBBS BMedSc FRACS MD
ACKNOWLEDGEMENTS

The following, encouraging words of Cowper are made more poignant by knowing they were written in 1699; his attitude has been a stimulus to the production of this thesis, and I quote:

"...there remain many considerable discoveries to be made....Of this the discovery of two glands....may be an instance....This may encourage us not to despond, if we do not find all enquiries attended by discoveries."

This study commenced when working as Senior Registrar to the Urology Unit in the Hospital for Sick Children, Great Ormond Street, London. Under the guidance and influence of Phillip Ransley and Patrick Duffy I set up a video recording system for urethral endoscopy; the ability to easily watch the procedure enabled me to be much more closely involved in the management of boys with congenital posterior urethral obstruction - for the first time I was able to see both the anatomy and the intervention. The tremendous resource of Great Ormond Street enabled this work to gain much importance, and the "thinking surgeon" approach of Phillip Ransley was a great stimulus. Discovery of the library of the Royal Society of Medicine allowed me to find and explore the many early works on congenital urethral obstruction.

I would like to pay tribute to the contribution of Hugh Hampton Young for his publications in the 1919 and 1929; without his contribution my work would not have been possible.

On arrival in Adelaide the theatre staff were willing to assist with the video taping which has been the centre-point of the study. The Urology Registrars and Fellows have also assisted greatly. Foundation studio staff have tolerated my many urgent requests for videos, prints and slides which has allowed for effective dissemination of the gained knowledge.

Neomedics and the parents of Jack Baker have assisted materially, with donations which have provided for the reviewing of videos, and production of the publications. The Adelaide Children's Hospital Research Foundation provided the money for purchase of computer equipment and the Kidney Foundation contributed by funding a summer scholarship which enabled Darrell Goh to assist with a further review of the video tapes.

Neil McMullin and Andrew Barker assisted with the collection of the data on renal transplant outcome in lower urinary tract obstruction patients, information which was provided from the ANZDATA registry for dialysis and transplantation. Doctor Toni Wilson helped with the co-ordination of the blood tests and literature retrieval for the boys with urethritis.

Brigid, my wife, has been a principle contributor to this thesis; the many evenings lost in a library, at meetings presenting the results, preparing manuscripts for publication or reviewing videos have been accepted graciously: this work is as much a result of her efforts as mine.
DEDICATION

To my mother and father for their encouragement and to my children as an example.
CONTENTS

CERTIFICATION
ACKNOWLEDGEMENTS
DEDICATION
CONTENTS 5

SUMMARY 7

INTRODUCTION 9

NORMAL ENDOSCOPIC URETHRAL ANATOMY

Introduction 11
Endoscopic Data 11
Discussion 13

MARION'S DISEASE

Introduction 14
Case Report 14
Discussion 18

CONGENITAL POSTERIOR URETHRAL OBSTRUCTION

History of Posterior Urethral Obstruction 20

Great Ormond Street Hospital Patients
Introduction 35
Prospective Study 35
Retrospective Study 41

Adelaide Children's Hospital Patients
Introduction 42
Materials and Methods 43
Results
Obstructive Membranes 44
Moderate Membranes 73
Minimal membranes 77
Position of the External Sphincter 80

Discussion
Classification 84
Embryology 90
Variable Expression 93
Position of the External Sphincter 94
SUMMARY

New technology for the examination of the lower urinary tract has resulted in higher quality information being available for study of congenital anomalies of the male urethra. The standard of the radiological images and the improved procurement techniques, together with the ability to easily view and record the urethra endoscopically, have allowed our understanding of urethral pathology to be reconsidered. Also, the availability of sophisticated perineal ultrasound, allows for a pre-emptive diagnosis of bladder neck and urethral obstruction so that an antegrade cystourethrogram can be appropriately performed, allowing the urethra to be viewed before any alteration of the anatomy. Combined with modifications of the conduct of endoscopy, recordings of the undisturbed anatomy have been able to be produced. This new information has indicated the need for re-evaluation of the method of dissection of the autopsy specimens and the need for new interpretations of the data; the development of new concepts has also been aided by the prenatal diagnosis of urinary outflow obstructive disease and the earlier presentation for treatment of the post-natal cases, which has strengthened the evidence of the congenital nature of the pathology.

All of the diseases studied in this thesis have been reassessed because of the facility to review video recordings of the pathology, while reviewing the cystourethograms, and the opportunity to compare the findings in the abnormal urethra with those of a large number of prospectively recorded normal urethras.

With the background of data collected in London, pertaining to obstruction in the posterior urethra, the video recording system was established for all my cases in Adelaide. The information obtained has provided proof of a case of primary bladder neck obstruction, further insight into classification of obstruction of the posterior urethra, variability of the degree of obstruction from case to case and the
relationship of the external sphincter to the obstructive membrane. Pathology of the more distal urethra has been differentiated from that of the posterior urethra, with the adoption of the term, Cobb's collar. The association of Cobb's collar to cystic lesions of Cowper's glands (syringoceles) has been assessed and a modified concept presented, suggesting that it is Cobb's collar, rather than a flat plate external sphincter, that syringoceles are closely related to.

The information on the outcome of transplantation in the urethral obstruction patients pertains to the secondary effects of congenital urethral obstruction. The detailed investigation of the cases of urethritis was undertaken as part of the study, to highlight the need to differentiate between congenital and acquired urethral pathology.

The clinical significance of the work is in the conceptual relocation of the proximal extent of the external sphincter in relation to syringoceles and posterior urethral obstruction, a new approach to intervention for urethritis in adolescents and raising concern about the long-term impact of outlet obstruction on bladder function.

From the academic perspective, the data provide new information on which to base embryological studies, plus, hopefully, a stimulus to others to undertake meticulous video recording to allow the hypotheses to be further tested.
INTRODUCTION

The currently accepted interpretation of urethral anatomy and pathology in boys is based on studies from the first half of this century, most notably from Hugh Hampton Young and his colleagues [1,2]. Surprisingly, no reports of systematic, urethral video-recording have been presented prior to publications from this thesis.

The strength of this work comes from the ability to review the endoscopic video tapes after subsequent cases have been identified. Thus, new cases of the same pathology were able to be compared directly with the early boys; those thought to have a separate disease were also able to be viewed in the one sitting. The set-up was, and needed to be, facilitated by a prospectively established data collection process, which was revamped after further video review. It was thus that the incidence of the bulbar urethral narrowing (Cobb’s collar) was discovered after the pathology became obvious from more notably obstructed cases. This group comparison has allowed a wealth of knowledge to be gained from a relatively small number of cases. Also, the recording of the first entry into the urethra, often without prior intervention, has also allowed for a different perspective to be gained, one which has been suggested by others [3-5]. However, their arguments have not gained general support, perhaps because of the lack of video recorded evidence. The ability to review the endoscopic recording with the urethrograms, when new questions arise, has also helped address the concerns of the sceptics.

Each segment of video was viewed at least twice, once at the time of the original endoscopy and a second time when the data were entered onto the computer, soon after the recording. Often previously unrecognised subtleties were noted at that stage. Subsequently, parents were often given the opportunity to view the video, thus I saw the video for the third time. In those cases where the pathology was of interest to the radiologists, the video was used for demonstration at a Hospital Nephro-urology Radiology conference. Many of the cases with a urethral anomaly
were reviewed on a number of occasions, some more than twenty times. The Cobb's collar and posterior urethral obstruction cases have also been shown to groups of Paediatric Urologists, to explore alternative perspectives and to act as a stimulus to improving the technique of information collection. In fact, the method of recording remained relatively unchanged over the five years of compilation in Adelaide.

The case of bladder neck obstruction, the discussion of the effect of outlet obstruction on renal transplantation and the dissertation on urethritis, while slightly peripheral to the central theme of the thesis, reflects the value of carefully considered and recorded information in the management of the lower urinary tract of boys, the secondary effect of bladder outlet obstruction and the importance of excluding acquired disease.
NORMAL ENDOSCOPIC URETHRAL ANATOMY

Introduction

Data on the normal anatomy of the male urethra has been collected from autopsy dissections of the fetus, boys and men [6-9], as well as from endoscopic [10,11] and radiological studies [12-15]. The data collected for this thesis was from boys who were found to have normal anatomy during an endoscopy for a wide variety of indications.

The urethra was considered to be normal if the sphincter could be seen to come to rest at the lower edge of the verumontanum when the bladder was empty. This positioning of the sphincter could obviously not be confirmed histologically, but it concurs with the endoscopic descriptions of others [6] and with the dissections of the fetus and boys of varying ages. A condensation of fibres at the lower end of the prostate or a physiologically more active region of muscle at the cranial end of the sphincter tube would concur with the endoscopic flat plate of the external sphincter, and be consistent with the anatomical dissections. The greater degree of activity of the muscle fibres located just beyond the prostate may be similar to the lower end sphincter in the oesophagus (Fig.1).

Endoscopic Data

Over the five year period in which patients were found to have the obstructive, syringocele and urethritis pathology described below, a further 119 boys had urethral endoscopy. The videos were reviewed to identify the anatomy of the posterior urethra, noting particularly the relationship between the verumontanum and the external sphincter; for the assessment of this part of the anatomy, the boys with urethritis were included as normal (as the posterior urethral anatomy was
normal in all). The sphincter (Fig. 1) was able to be viewed in all boys where an adequate recording of the posterior urethra had been obtained, which was in 87 of the 126 cases. Prominent folds extending from the distal end of the verumontanum were also seen as shown in Figure 1B.

Figure 1A: The normal urethral sphincter as seen endoscopically, contracting onto the distal end of the verumontanum.

Figure 1B: The normal urethral sphincter overlying the distal end of the verumontanum with minor folds attached to the crista urethralis.
The number of cases with an inadequate recording of the posterior urethra relates to the early patients only having a recording of the urethra when pathology was expected. All patients were always examined with the first insertion of the endoscope, but not all were recorded. For boys who had more than one cystoscopy, but no recording of the urethra during one of the cystoscopies, other studies of the same patient were reviewed.

Discussion

Oelrich and Hutch et al. performed detailed dissections of the urethral sphincter at various ages, concluding that the sphincter is a continuous tube of muscle from the bladder to the perineal membrane, with a relative condensation of fibres caused by the growth of the prostate gland [6,8]; an increased number of fibres at the distal end of this tube of muscle may account for the appearance of the muscular Cobb's collar in the bulbar urethra [16].

Stephens [17,18], Gibbons et al. [19] and Currarino [14] suggested, from cystogram studies, that the muscular, variable constriction of the bulb is due to contraction of the separate striated muscle of the bulbospongiosus contraction. However, the anatomical dissections of Hutch et al. [8] and Olerich [6], suggest that the continuous tube of striated muscle forms a lower "muscle complex", not too dissimilar to the muscle complex associated with the lower bowel. The results of these studies are supported by the findings of Bradford Young [20] and the cystograms in this thesis. Other authors highlight the predominance of the striated muscle fibres in the anterior wall of the urethra of the most physiologically active component of the external sphincter (the endoscopically identified sphincter), which appears to be positioned at the distal end of the verumontanum [8,9]. This would fit with the endoscopic impression from the 87 normal endoscopies in this study.
MARION'S DISEASE

Introduction

Marion (1933) was the first to describe the condition of primary muscular hypertrophy of the bladder neck leading to obstructive uropathy, in a group of patients who had died of complications of renal failure [21]. Although bladder neck hypertrophy can be seen with more distal obstruction, it is felt that these patients represent a different disease. The condition has since been described by others [22-25], but, until this case, a patient with prenatal hydronephrosis and primary bladder neck obstruction had not been reported. The diagnosis of muscular hypertrophy with obstruction was supported by perineal ultrasound (US), which showed a prominent bladder neck, and a renal US which demonstrated severe hydronephrosis. Also, an antegrade cystogram, plus suprapubic and urethral endoscopy, showed a narrow lumen through the bladder neck and obstructive changes in the bladder, and a biopsy of the bladder neck was consistent with muscular hypertrophy.

Case Report

GP (UR: 9111523) was born in October 1991 at 34 week gestation. His first antenatal US at 27 weeks showed a distended bladder which increased in size on subsequent scans, and minor hydronephrosis which progressed to bilateral severe hydronephrosis, with parenchymal thinning, postnatally (Fig.2). A perineal US demonstrated the muscular hypertrophy consistent with bladder neck obstruction (Fig.3). A urethrogram performed through the suprapubic catheter showed high grade vesicoureteric reflux (VUR) into bilateral megaureters and a very narrow lumen through the bladder neck (Fig.4). A nuclear medicine scan indicated that the function of the right kidney was reduced to 25% of overall renal function, but total renal function was normal.
Cystourethroscopy was attempted with a 7.5FG scope via the penis, without success, as the bladder neck was impassable. A 9.5FG scope was then passed along the suprapubic tract, identifying a prominent bladder neck, particularly the posterior portion, which protruded into a trabeculated bladder (Fig.5). A guide wire was passed through the bladder and into the urethra, which allowed dilatation of the bladder neck, after which an 8FG urethral catheter could be inserted.

A follow-up cystogram showed the bladder neck to be persistently narrow. Therefore, the bladder neck was incised, biopsied and closed transversely at the time of performing a vesicostomy. Repair of a prolapsed vesicostomy failed to prevent a decline in the infant's renal status on nuclear scanning, leading to repeat cystoscopy with a view to treating his persistent VUR and closing his vesicostomy. His proximal urethra and bladder neck easily admitted a 9.5FG cystoscope, and his bladder neck was satisfactory when viewed via the vesicostomy (Fig.6). Hence, bilateral tapering ureteric reimplants were performed and the vesicostomy was closed. It is note-worthy that the cystogram through the vesicostomy showed some improvement in the bladder neck diameter, but not to the degree seen endoscopically. Post-operatively he was voiding with a good urinary stream. Ultrasound showed decreased pelvocalyceal dilatation, he continues to void well and a nuclear medicine study indicated that his renal function has been stable.
Figure 2: An early postnatal ultrasound showing marked hydronephrosis and thin parenchyma of the right kidney.

Figure 3: A perineal ultrasound soon after birth showing the hypertrophied bladder neck projecting into the bladder. The image is orientated the same way as the cystourethrogram in Fig.4.

Figure 4: A suprapubic cystogram shows a virtually nonexistent proximal urethral lumen prior to intervention.
Figure 5: An endoscopic view of the bladder neck reveals the narrow firm bladder neck, with the greater prominence posteriorly (lower edge of picture).

Figure 6: A suprapubic cystoscopic view of the bladder neck several months after vesicourethroplasty. The bladder neck is notably wider than previously.
Discussion

A definitive diagnosis of primary bladder neck obstruction is difficult to make, particularly as there has been much confusion in the past as to what constitutes bladder neck hypertrophy of both a primary or secondary nature [22,23,26,27]. Secondary bladder neck prominence is seen with distal obstruction and with a neuropathic bladder, but the degree of true obstruction at the bladder neck is often uncertain. In patients with detrusor-sphincter dyssynergia, the apparent functional obstruction is part of the complex interaction of the muscular components of the bladder which most would regard as secondary bladder neck pathology. Previous authors have advocated bladder neck surgery for the management of VUR and functional outlet obstruction, an indication which has not been substantiated by more recent reports [23,27], and has been disputed by others [28].

Primary bladder neck obstruction was first described by Marion, who examined post-mortem specimens of 77 patients who died of obstructive uropathy [21]. He based the diagnosis on either the failure of the bladder neck to open during micturition or the inability to pass a sound into the bladder, in the absence of dilatation of the posterior urethra. Bodian [24] reported five autopsy cases in males and found a predominance of fibroelastic fibres. Cross-sections of the specimens are given in his paper, which differ from this case because of the presence of elongation of the prostatic urethra; significantly "no mechanical obstruction" was found in any of the specimens, suggesting that the changes seen may have been secondary to high grade VUR. McDonald et al. [26] appears to have reported distal obstruction with secondary bladder neck hypertrophy. Andreassen presents more convincing evidence for congenital, primary bladder neck obstruction with a series of 6 cases, three of whom had a thickened bladder neck "resembling a pyloric stenosis" [22], as did this patient (Fig.3+5). However, two of his cases may have had detrusor-sphincter dyssynergia and one was a patient with a neuropathic bladder and spina bifida.
Adults with urodynamically demonstrated functional bladder neck obstruction [27,29] probably represent a different disease to the mechanical obstruction seen in children. In children, the diagnosis is made by radiologic and endoscopic demonstration of narrowing of bladder neck with failure of funnelling of the posterior urethra on micturition cystourethrogram (MCU), and the presence of a circumferential ridge of tissue protruding into the bladder on cystoscopy and perineal US. Williams suggests that bladder trabeculation must also be present for a definitive diagnosis [30], a finding in this case.

The pathology of the mechanical obstruction is not certain; Bodian thought it was due to fibro-elastosis [24], Young considered the fibro-elastosis secondary to infection [31], which Presman also described as fibro-muscular hyperplasia in four cases [32], but of the posterior urethra rather than the bladder neck. In the boy presented above, the bladder neck biopsy showed bundles of hypertrophied smooth muscle and a small amount of connective tissue. This is consistent with the observations of Kaplan and King [23], who described it as the admixture of various amounts of normal or hypertrophied smooth muscle, fibrosis and elastic tissue.

Primary bladder neck obstruction can be treated effectively either by open operation or endoscopically. Andersen advocates endoscopic incision of the posterior bladder neck [29]. Kaplan and King [23], and Bradford Young [20] describe an open Y-V vesicourethroplasty, and mention removal of a wedge of the posterior bladder neck as an alternative. I chose to excise a wedge of the posterior bladder neck for the purpose of obtaining a biopsy and closed the defect transversely, thus widening the extremely narrow bladder outlet.
CONGENITAL POSTERIOR URETHRAL OBSTRUCTION

History of Posterior Urethral Obstruction

The earliest description of obstruction of the posterior urethra was by Morgagni in 1717 [33]. His and subsequent descriptions in the 19th century were based on post-mortem dissections, the first of which was included in Langenbeck's monograph on stone disease, in 1802 [34]. Velpeau, in 1832, described folds in the posterior urethra which resembled valves, and mentioned the possibility of their causing obstruction to the passage of a catheter [35]. Over the following 85 years there were 27 reports identified in the literature, most of which have been referred to in the pivotal papers by Young et al. in 1919 [1] and Hinman and Kutzmann in 1925 [36]; however, not all reports were included in these papers and not all cases included would have qualified if the reports were being prepared today. The main features of each of the reports are as follows:

Budd, in 1840, presented the case of a 16 year old sailor who presented in a moribund state, died and was found to have ureters as large as intestine and a dilated, thickened bladder; in the upper wall of the membranous urethra was a fold of membrane similar to the valve in a vein [37]. In 1847, Bednar performed an autopsy on a 12 day old premature infant who died after 5 days of urinary retention. The lower end of the verumontanum was found to divide into two concave folds facing the bladder, the kidneys were atrophic and hydronephrotic. The next report identified, comes from the minutes of a meeting at which Godart presented the 11th case to the Société Anatomique, in 1854. The entire report read, "Enfin le même présentateur met sous les yeux de la Société la pièce suivante: Dans le canal de l'urètre, sur un des côtés du verumontanum, on voit une valvule membraneuse fine, délicate, rappelant par sa forme un nid de pigeon, les valvules sigmoïdes, ou encore les valvules de Tarin. La portion libre regarde vers le col de la vessie; son bord adhérent est tourné en avant. Aucun auteur n'a parlé d'une semblable valvule"; that is, the autopsy showed a membranous valve at the level of the verumontanum in the posterior urethra, in
the shape of the nest of a pigeon, with the free edge toward the vesical neck. Although the description is flamboyant, the author leaves you in little doubt about the nature of the pathology. Unfortunately, no illustrations were included.

Picard’s case, presented in 1855 [38], was a 40 year old brewer who became unconscious and convulsed while undressing, and died shortly after. Before his demise, he was noted to have a distended bladder; urethral intubation was unsuccessful until a smaller catheter was used. At autopsy the resistance to catheterisation was found to be a valvular obstruction below the verumontanum.

Jarjavay’s recording in his monograph in 1856 [39], likened the obstruction to the iris of an eye, after identifying "that seen in the anterior portion of the pars prostatica, a circular membrane with its periphery attached to the urethral wall, having in its centre an opening 3 mm in diameter". However, it was not until Tolmatschew presented an autopsy which showed two valves just below the verumontanum that the first attempt to classify posterior urethral obstruction was made. Although Tolmatschew did not provide any clinical history or examination, he gave a full description of the posterior urethral findings and reviewed the literature up to 1870 [40].

Lowsley, in 1914, refers to a case in which Ebert "punctured the obstructing membrane in a similar case", around 1890 [41] and, in 1891, Eigenbrodt [42] reported a 54 year old man who was found at autopsy to have bladder outflow obstruction from a flap of tissue at the bladder neck, similar to the case in which endoscopy was first mentioned by Poppert, in 1892 [43]. The first of these two reports was included in the list of cases by Hinman and Kutzmann, but not mentioned by Young et al. 1919 [1]. Eigenbrodt's case was mentioned, for exclusion, in 1929 [2]. Neither of the reports appear to be of a definite posterior urethral obstruction. However, Poppert's patient, a 24 year old who had difficulty voiding from childhood, was almost certainly suffering from a congenital lesion, the nature of which was confirmed at autopsy.
Interestingly, the 1893 report of Balantyne [44] is not mentioned in either of the summary papers of Young et al. [1] or Hinman and Kutzmann [36]. He described a stillborn infant with a bladder which was distended with clear, slightly straw-coloured urine, with a fundus that reached the umbilicus, and beyond, on either side of the midline. "In the urethra a membranous septum existed at a point in front of the level of the symphysis pubis; but whether this was completely impervious or not must be left doubtful, as it was injured during dissection." The peritoneal cavity appeared to contain ascites. Thus the first, definitive case of congenital obstruction of the posterior urethra was presented. The two reports in 1895 were Christen's case of a valvular stricture in the prostatic region following on an hypertrophy of the crista [43], and Porak's stillborn infant with dystocia from a large, thin walled bladder of 120 ml [45]. The description of the posterior urethra was similar to that of Godart; "in the posterior urethra was a mucous fold in the shape of a valve with its concavity to the bladder and extended into the membranous urethra, terminating at the veru".

A significant development at this stage was the advent of radiography in 1895, with the subsequent use of the cystogram.

The evidence for a malformation causing the abnormality was again documented in the final two cases reported in the 19th century. The first was in 1896 when Schlagenhauer's patient, who died at birth, was identified to have a valvular obstruction at the lower end of the verumontanum [46]. Interestingly, in contrast to Picard's case, the obstruction could be passed by a large sound, but not a fine probe. The second was a boy who died at birth in 1898 [47], in whom a delicate valve-like structure was noted at the lower end of the verumontanum, which was attached to the floor of the urethra and was associated with bilateral hydroureteronephrosis. The first two cases reported in the 20th century were published by Fuchs in 1900, one a five month old and the other a stillborn child, both with a valvular obstruction below the verumontanum [48].
The turn of the century saw a change in the nature of the reports on posterior urethral obstruction; still a number of case reports were presented, but authors became more intent on understanding the aetiology, presentation and management. For instance, Bazy (1903), in addition to presenting six cases, went on to give a dissertation on the possible embryology [49].

In 1904, Lindemann [50] reviewed three children one each of 4 months, 4 years and 5.5 years. All three had a valvular obstruction emanating from the anterior end of the verumontanum, plus bilateral hydroureteronephrosis. One of these boys had a systemic infective illness, as did the case of Morton, in 1904, [51] and Wilckens, in 1910 [52].

Thompson, in 1907, [53] expressed a similar view to Bazy, but used different terminology. He stated that the septum had a central aperture and was at the junction of the urethral orifice of the bladder and the first portion of the urethra. He wrote, "presumably this septum must be the structure formed by the cloacal hypoblast and the invaginating perineal epiblast analogous to the anal canal and hymeneal membranes [53]."

Anaemia and convulsions were not uncommon manifestations of the disease at this stage of understanding, as recorded by Fletcher (1908) [54] and Wilckens (1910) [52].

The pathology was generally reported as a valve-like obstruction in the posterior urethra, with occasional variations in the description such as a "three membranous valve-like obstruction" [54] or "six folds below the verumontanum" [50]. Lederer provided two views of the autopsy specimen of his 11 year old patient, one, a view from above and the other, the urethra split in the anterior midline, concluding the abnormality to be a "semicircular diaphragmatic obstruction below the verumontanum in the posterior urethra" [55].
The first American report, and possible the first clinical diagnosis, was the case of Knox and Sprunt in 1912 [56]. Their patient presented to the paediatric service of the John Hopkins Hospital in Baltimore, at which Hugh Hampton Young was working.

The first large series of patients reported in the 20th century, a group of 19 cases of congenital stricture, was by Riedel (1913), quoted by Lowsley in 1914 [41]. The other two reports in 1913 were of young children who had symptoms from birth, one, a five year old with a urinary tract infection (Heinecke [57]), the other, a four week old with continued dribbling found to have a fibrous stricture in the posterior urethra on autopsy (Jordan [58]). Whether these were part of the same disease is uncertain.

The inclusion of Iverson's case from 1914 seems dubious. He treated an 85 year old man for haematuria, difficulty micturating and a distended bladder [59]. Iverson had been unable to pass a catheter because of what was thought to be a false passage: at autopsy a valve-like structure was found at the lower end of the verumontanum, which on the evidence may well have been due to a false passage rather than a congenital lesion. Of note, the case was included in the each of the presentations by Young et al. [1,2] and that by Hinman and Kurtzmann [36], although the patient was 85 years old, not 15 years old as given in the table in the 1919 paper.

Lowsley (1914), in identifying a newborn boy who died of fever and abdominal distension, indicated the early contribution of Young's thinking when discussing the findings. "...the fact that the entire structure is more or less dome shaped, make the term "diaphragm"(!) suggested by Dr Hugh Young, seem most appropriate in referring to this anomaly" [41]. The final publication before the seminal paper by Young et al. was a report of a 5.5 year old and a 7.5 year old by Artus in 1917 [60], the foundation year of the Journal of Urology.

Up until 1919, 24 authors were credited with making case comments or presentations
Congenital Posterior Urethral Obstruction - History

related to posterior urethral obstruction, according to Young et al. [1], whereas 26 were retrieved by the Hinman and Kutzmann paper [36]; five were included in the classification in one study and not the other. I was able to find three others from alternative sources. The lack of access to Medline and computers would easily explain the discrepancies.

The pivotal paper, written by Hugh Hampton Young [61] and his co-authors William A. Frontz and John C. Baldwin in 1919 [1], was read at the June meeting of the Association of Genito-Urinary Surgeons in Atlantic City. It was a report of 12 cases treated at Johns Hopkins Hospital, Baltimore and the six part classification was reiterated in the Young and McKay paper in 1929, which added nine new patients [2].

The Young et al. cases ranged in age from 11 days to 42 years, with only five being of 12 months old or less. Five cases had an autopsy diagnosis, five had a clinical diagnosis and three (including one that later had a post-mortem) had an endoscopy. Two of the diagnoses were with the finger passed through the bladder neck. The 1919 paper was the first to describe the endoscopic diagnosis of urethral "valves", and to develop a transurethral "punch" for treating the obstruction. The authors gave a great deal of detail for each of the patients [2].

Until Young made an endoscopic diagnosis, posterior urethral valves were identified either at autopsy or by the passage of sounds [49]. Obviously, the latter did not help to elucidate the anatomy of urethral obstruction, but less apparent is the inaccuracy of the method used for post-mortem dissection. A number of studies have subsequently looked at the anatomy, embryology and the clinical outcome, including the long-term effect on the bladder, after the obstruction has been relieved.

Hinman and Kutzmann [36] reviewed the following cases which were reported after the paper by Young et al. in 1919. Silverberg, in 1920, presented a case which had been investigated both radiologically and endoscopically, following a urinary tract infection [62].
A year later Randall was the first to perform an endoscopic resection. Both the five and 16 year old boys treated by fulguration did well [63] as did two cases treated by Glingar in 1922 [64].

Further cases were reported in third decade of the 20th century including those by Martin (1922) [65], Martinson and Reuben (1923) [66], Schmidt (1923) [67], Hausmann (1924) [68], Bronner (1924) [69] and a case of Type II by Ehrich (1925) [70]. In all, Hinman and Kutzmann [36] added six of their own and collated 50 cases from the literature, 14 of whom were greater than 10 years old. Two, in fact, had a flap of tissue in the bladder, and therefore should not have been included (Eigenbrodt 1891, Schmidt 1923). The six new cases presented by Hinman and Kutzmann ranged in age from two to 57 years. Inadequacies of the paper were the inclusion of an 18 year old who did not have any obstructing lesion seen endoscopically and most of the cystograms were of poor quality and, therefore, of little value in precise evaluation of the anatomy. It should be remembered that the lighting for, and size of, cystoscopes has improved dramatically in recent years.

In 1931, Campbell reported 18 cases, in whom radiography was usually performed and cystostomy drainage was common. However, four refused treatment (outcome unknown), six died, and only eight had symptomatic relief. The treatment options used were Young's miniature punch, fulguration or passage of a metal sound [71].

Lowsley and Kirwin added three new cases and reviewed the literature, identifying a total of 130 cases up to 1934 [72]. Authors not mentioned in previous compilations include Poynton and Sheldon (1927), Day and Vivian (1927), Morris and McCrea (1928), Kretschmer (1929), Addison (1929), Bigler (1929), Ormond (1929), Schacht (1930), Chadwick et al. (1930), Hullsick (1930), and Browne (1930).

Significant improvement in the understanding of congenital posterior urethral obstruction
came with refinement of the cystogram. In 1955, Stephens presented the detailed findings of the normal and abnormal urethra during an MCU, highlighting the importance of the oblique view in assessing the anatomy [17]. His opinion was supported in the study by Griesbach et al. in 1959 [73].

New techniques have allowed for the avoidance of urinary diversion, with the primary focus tending toward endoscopic ablation of the obstruction, using newly developed small cystoscopes. While awaiting the advent of these instruments, Johnston used an auriscope through the perineum, facilitating the use of electrocautery and reducing the risk of urethral stricture formation [74]. Duckett and Snow [75] also advocated the use of primary ablative surgery, but used a resectoscope loop converted to a hook, stressing the need to avoid stricture formation, but only if progressive renal deterioration occurs should upper tract diversion be performed. Other per urethral interventions include the use of the Fogarty catheter or an insulated diathermy hook developed by Williams et al. (1973) [76]; a technique further developed by Whitaker and Sherwood [77]. The move away from ureterostomy and vesicostomy has not been universal as Kumar et al., reported the use in 3 of 32 cases in 1972 [78], and Kendall and Karafin, although advocating primary treatment of the urethral obstruction, preferably via the perineum, supported the use of ureterostomy if the upper tracts did not deflate [79]. This latter view was shared by Duckett and Snow [75], whereas, Warshaw, in 1985, [80] reported the use of cutaneous ureterostomy or pyelostomy in 13 of 22 cases. In the presence of upper tract dilatation, after ablation of the urethral obstruction, simple elevation of the urine collection device seemed to improve the ureteric outflow in the patients treated in Adelaide.

A further change in the surgical treatment has been the indication for nephrectomy. Warshaw performed a nephrectomy in six of his 22 cases [80], expecting to remove a non-functioning unit to prevent infection and hypertension, as was advocated by Hoover and Duckett [81]. It became increasingly recognised that the subsequent deterioration of the remaining contralateral kidney can occur. Also, bladder dysfunction can contribute to
Congenital Posterior Urethral Obstruction - History

incontinence [82,83], late renal failure [83,84] and failure of subsequent renal transplant. This understanding has led to ureter preservation, which enables ureterocystoplasty to be a subsequent alternative means of bladder augmentation [85-90], particularly when the renal dysplasia is associated with a huge refluxing megaureter as occurs in the VURD (vesicoureteric reflux and dysplasia) syndrome. This phenomenon may protect the contralateral side from renal injury [81,91]. The development of a large congenital bladder diverticulum and urinary ascites, particularly prenatal ascites, may also protect the upper tracts [91]. The recognition of these phenomena has been important in the understanding of the natural history of the condition and the optimum care of the boys.

Over recent times, the survival rate has continued to improve and the nature of the presentation has changed from the complications of renal disease, to complications of bladder disease, through to symptoms of bladder disease. Now, in fact, most Australian cases are identified as an incidental finding on prenatal US.

By comparison, in 1966, Ellis et al. reported 29 cases in the Columbus Children's Hospital, treated between 1947 and 1965, or just over two per year. Only 14 others had urinary symptoms. All those older than six months lived, but 14 died [92]. Then, in 1973, Williams et al. published the results of 206 cases managed from 1951 to 1970: the improving outcome is highlighted by a fall in death rate from 36% to zero in the final 21 cases [76]. The younger presentation meant that the disease was being detected earlier, but in patients who might be harder to manage because of their young age. Despite the younger age of presentation and consequent greater technical difficulty, Churchill et al. found the mortality in the 70's improved with time in most units [83], which was the individual unit experience of Cass and Stephens [93]. They found that from 1950 and 1961, the death rate was 13.0% over one year, 70.1% under one year, but 77.8% if less than one month of age. In the decade after 1962, the mortality was zero for the over one year old group and only 21% under one year of age. Results have continued to improve for live born children.
An interest in the genetic aspects of congenital urethral obstruction has become more prevalent, with reports of sibling and twin cases [94,95]. Inheritance across successive generations has also been recorded, but the role of genetic factors is poorly understood. As will be discussed later, four of the 29 boys with posterior urethral obstruction, in Adelaide, came from a twin pregnancy.

A significant change in the patient profile occurred in the 80's, noted in the paper by Scott in 1985 [96]. He reported his experience with 46 cases in whom two had a fetal diagnosis, 52% were diagnosed in the first three months of life and only two had upper tract diversion.

Fetal diagnosis is now frequent, as indicated by the findings for the Adelaide patients: over the five year study period, 12 were diagnosed antenatally as having probable posterior urethral obstruction. Two fetuses were terminated and found to have renal dysplasia at 23 and 20 weeks, three were delivered prematurely or allowed to go to term and died of pulmonary hypoplasia, with poor renal tissue at autopsy. Of those who survived, two are likely to require renal transplantation in the first decade, but the others, including the twin of 29 weeks gestation, have a good prognosis. Five had primary ablation, one was managed by urethral catheterization and the other had a vescicostomy formed, which was only used because the child was 29 weeks gestation and had hypospadias. No ureterostomies were required and three ureterocystoplasties have been performed. Kendall and Karafin, in 1975, made no mention of prenatal diagnosis, although 24 of their 34 cases were less than 4 months old at the time of presentation [79].

Paediatric urology has advanced significantly since the discussion in the Royal Society in 1835, about whether the fetal kidney is able to make urine [97]. It is now known that fetal urine production commences in the 14th or 15th week of gestation [10]. Because of this urine production, the detection of renal tract anomalies is common, representing half the
Congenital Posterior Urethral Obstruction - History

antenatally detected abnormalities and thus is seen in 0.1-0.25% of pregnancies [98]. Despite our improved understanding, classification of fetal hydronephrosis is not yet agreed upon, although the possible relationship to urethral obstruction is well recognised [98]. A fetal renal pelvic diameter of >15 mm after 20 weeks gestation [99], and an attempt to grade parenchymal thickness [100], the pelvic to parenchymal ratio [101], and the kidney to abdominal wall circumference have all been suggested as possible standards indicating significant pathology. The majority of first scans are performed between 17 and 19 weeks gestation, and therefore guide-lines for pelvic dilatation are more valuable at this gestation. These have not been well established, possibly because the majority of those with minor dilatation early in pregnancy resolve spontaneously. Mandell et al. have suggested the diameters of 5 mm at 15-20 weeks, 8 mm at 20-30 weeks and 10 mm thereafter, are kidneys worthy of ultrasound review. Using these criteria, 20-37% of this group require surgery [102,103]. Importantly, 30% of those with urethral obstruction, or other significant obstructive renal tract pathology, have a normal scan before 20 weeks gestation [104]. Never-the-less, ureteric dilatation is always significant, and suggestive of severe VUR, or obstruction of either the ureter or urethra [105].

Echogenicity is a further factor used to illucidate the significance of fetal hydronephrosis. However, establishing a standard for echogenicity is difficult, principally because the fetal renal parenchyma is relatively echogenic compared to children and adults. Estroff et al., however, found that 80% of kidneys that were more echogenic than the liver had abnormal function at birth [106]. A more specific sign of renal dysplasia is the presence of parenchymal cysts, but less than half of the fetuses with dysplastic kidneys have cysts identified on ultrasound, and early in gestation, cysts are less likely to be detected in subsequently proven renal dysplasia [107].

The most conclusive study of fetal renal deterioration is a series of scans of the same fetus which show progressive changes in the echogenicity, the development of cysts, or a thinning of the renal mantel over time.
Prenatal intervention has developed from the ability to obtain information on the renal tract of the fetus. The main benefit of prenatal identification of the abnormal renal tract has been the improved understanding of renal tract pathology in children; the anticipated benefits of prenatal intervention for nephron preservation have not as yet been widely achieved [108]. This is mainly because of limitations in identifying those who will benefit from intervention, the short-comings of stenting techniques, and the risks of open fetal surgery. However, animal experiments have shown that obstruction of the fetal urinary tract at either the ureteric or bladder neck level can cause renal dysplasia if instigated sufficiently early in gestation [109,110], and the effect of obstruction on renal development can be reversed by relieving the obstruction by vesicostomy [111] or inserting a shunt [110]. The success in animals has validated attempts to consider intervention in the management of the obstructed fetal bladder outlet in humans [112,113]. Chromosomal analysis should always be performed where intervention is planned, or in the presence of a co-existing abnormality, where the risk of an aberration may be as high as 24% [114].

A number of factors need to be considered in selecting a fetus for intervention, after chromosomal anomalies and other congenital defects have been taken into account, including the number of possible causes of fetal renal tract dilatation. The importance of differentiating between many other pathologies can not be over stated. Firstly, as prenatal intervention has a maternal risk, the age and pregnancy history of the mother, the difficulty conceiving, the number of fetuses and the gestation are all important; the risks of fetal loss and chorioamnionitis [112,115] should be fully considered before proceeding. Also, intervention is unwarranted in transient obstruction, for VUR, in patients who would be more appropriately managed postnatally, or in those for whom the intervention would not improve the renal function outcome, such as unilateral disease or where irreversible dysplasia already exists, megacystis microcolon hypoperistalsis syndrome, cloacal anomalies.
An obstructive lesion is suggested by progressive pelvic dilatation with thinning of the parenchyma, a decrease in the amniotic fluid, urinary ascites or urinoma, and in the case of a dilated bladder, a prominent proximal urethra and poor bladder emptying. These features indicate that obstruction is probable; the question of prognosis depends on lung development and renal function potential. For lung growth to be adequate, the amniotic fluid has to be satisfactory early in pregnancy or lung hypoplasia will occur, even if the amniotic fluid has been improved subsequently [116]. Conversely, if intervention is instigated soon after the onset of oligohydramnios late in the pregnancy, the prognosis for both lung and renal function is good [117]. In fact, a decrease in amniotic fluid should be regarded as a relatively late sign and an adequate amniotic fluid should not be a contraindication to intervention.

The presence of bilateral renal cysts should preclude intervention, whereas a definite bladder outlet obstruction with initially normal kidneys and amniotic fluid, with progressive dilatation, but satisfactory fetal urine biochemistry, is the ideal case for draining the renal tract. Where the amniotic fluid volume is already markedly decreased and the urine biochemistry grossly abnormal, the prognosis is less favourable.

If the US changes suggest intervention may be appropriate, aspiration of the renal tract should pre-empt any definitive procedure. The fetus should be otherwise normal, and intervention should be expected to follow if appropriate. While performing the bladder aspiration, the volume, pressure and the response of the upper tract dilatation to the bladder emptying should be assessed [118]. Urine output can also be measured by leaving the needle in the bladder for a period of time. The fetus should be paralysed, and amniotic fluid should be collected for microprotein analysis and karyotyping.

Harrison's group looked at biochemical prognostic indicators in animals and then in patients [112], but did not provide data related to gestational age. Nicolini et al. have provided distribution tables of sodium for gestational age, showing that expected levels
decline with gestation and increase with renal damage [119]. They recorded the normal sodium range at 16 weeks gestation as 60-130 mmol/l and at 36 weeks, 10-60 mmol/l. The sodium, amniotic fluid, and ultrasound changes have been found to be more reliable than the urine creatinine, potassium and N-Acetyl-β-D-Glucosaminidase (NAG) in predicting the renal function outcome [108,112].

The microproteins β2- and α1- microglobulin have been recently explored, and are thought to be better predictors of renal function than the sodium level. There is normally a progressive decrease in these levels toward the end of pregnancy, as for sodium [120]. The microproteins are normally filtered through the glomerulus and resorbed in the proximal tubules. In the damaged kidney they are able to leak into the urine, and can then be measure in both the fetal urine and the amniotic fluid. The levels correlate well with fetal renal damage as the microproteins are unable to cross the placenta [108,121].

In general, a fetal urine sodium of <50 mmol/l and a β2-microglobulin of less the 2 mg/l are indicative of good renal function at the time of sampling. A urine sodium of >70 mmol/l and a β2-microglobulin of >15 mg/l are virtually always associated with poor renal outcome.

There are a number of fetal intervention alternatives available, including early delivery. After initial enthusiasm for inserting catheters into the fetal bladder to by-pass the urethral obstruction, the most likely current management is to await term delivery or arranging for a timely induction of labour.

Harrison et al. demonstrated the efficacy of percutaneously placed bladder catheters and forming a vesicostomy in sheep [110,111]. Due to the problems with percutaneous shunts in humans, such as incorrect placement, dislodgement and ineffective drainage [115], a fetoscopic approach with both stent insertion [122] and laser diathermy of the fetal abdominal wall [123] have been explored. The majority of human fetuses treated with
urinary tract diversion have had a percutaneous shunt inserted which, if required early in gestation, usually has to be replaced; a small number of cases have undergone a fetal vesicostomy [113]. As yet the prognosis for those thus treated has not been satisfactory [108], usually related to an inability to identify which case has reversible renal damage.

In the case of a fetus with cystic or highly echogenic kidneys in the early pregnancy, with little or no amniotic fluid, the prognosis is universally poor, and therefore, fetal bladder aspiration and treatment are not appropriate. Therefore, the focus for renal salvage may be more appropriately directed to the 30% with late onset ultrasound changes. In these, the lung development is normal and the renal parenchyma is probably initially normal; they require only short term urinary diversion or can be considered for early delivery after steroid stimulation for lung maturity.

The history of the evolution of the understanding of congenital obstruction of the urethra in the male, as for all diseases in medicine, is well worth studying as it gives insight into the nature of the material and its assessment, and gives us the background on which we build our knowledge. The poor quality of the radiology and the endoscopic equipment necessitates a relook at the pathology with our new instruments and understanding.
CONGENITAL POSTERIOR URETHRAL OBSTRUCTION
Great Ormond Street Hospital Patients

Introduction

Seven boys had cystourethroscopy specifically to assess the anatomy of their posterior urethral obstruction. These represent seven consecutive cases of posterior urethral "valves" treated at Great Ormond Street between July 1989 and July 1990. Five had a suprapubic cystostomy for initial bladder drainage, four without prior urethral instrumentation. The data were collected prospectively and the videos subsequently reviewed on multiple occasions, including after the Adelaide cases had been recruited.

Prospective Study

Case 1. M.K. showed moderately severe obstructive uropathy from 18 weeks gestation on prenatal US. However, the amniotic fluid volume was normal. He was born at term with a palpable bladder. An US was suggestive of urethral obstruction, so a suprapubic catheter was inserted on day two, after transfer to GOS Hospital. On endoscopy, a membranous obstruction was seen in the posterior urethra with a small defect posteriorly, consistent with the MCU findings. A Whitaker diathermy hook was used to split the membrane, producing two firm-edged "leaflets" which relieved the obstruction, as shown on a postoperative urethrogram.

Case 2. J.W., the first born of twins, was noted to have bilateral hydronephrosis and a distended bladder at 36 weeks gestation. He was transferred for urological care within 24 hours of birth when US confirmed the prenatal findings, leading to the insertion of a suprapubic catheter. An antegrade contrast study showed urethral obstruction, an abnormal bladder and left VUR (Fig.7A). At endoscopy, a membranous obstruction was seen, with a posterior defect approximately the diameter of a 3FG ureteric catheter (Fig.7B). A Whitaker hook was used to disrupt the membrane, following which the sphincter could be seen coming to rest part way along the "leaflets" (Fig.7C).
Figure 7A: The cystogram of Case 2: The typical changes of elongation and widening of the posterior urethra can be seen.

Figure 7B+C: The endoscopic findings of Case 2: The posterior defect is shown adjacent to a 3FG ureteric catheter (B), and the external sphincter can be seen coming to lie against the folds running down from the verumontanum (C).
Case 3. G.H. presented for treatment on the first day of life, with a prenatal diagnosis of bilateral hydronephrosis which was first detected at 18 weeks gestation. He emptied his bladder during an attempt to insert a suprapubic catheter, therefore, a 6FG feeding tube was inserted to fill the bladder and facilitate the insertion of the suprapubic tube. An MCU showed posterior urethral obstruction and a filling defect in the membranous urethra. Endoscopy demonstrated a punctured syringocele and a posterior urethral membrane with a posterior, midline defect adjacent to the verumontanum. A single passage of a Whitaker diathermy hook produced a "valve leaflet" appearance.

Case 4. C.M. was the product of a normal pregnancy and delivery, having one normal US at 16 weeks. Subsequently, he was noted to have haematuria during a febrile illness at the age of three weeks. Urinary infection was confirmed, and an US showed bilateral hydronephrosis and evidence of bladder outflow obstruction. He had a Creatinine (Cr) level of 185 mmol/l. An MCU via a suprapubic catheter showed obstruction of the posterior urethra (Fig.8A). Endoscopy showed a urethral membrane with a posterior 3 mm defect (Fig.8B). The membrane bulged markedly on bladder compression (Fig.8C+D). The lesion was diathermied with a Bugbee electrode, and three months later the remnants of the "valve leaflets" were resected. His renal function remained moderately impaired.

Figure 8A: The cystogram of Case 4, showing elongation and widening of the posterior urethra with a sudden change in calibre at the level of the obstruction.

Figure 8B: The endoscopic findings of Case 4: The posterior defect is shown adjacent to a Bugbee electrode.
Figure 8C+D: The endoscopic findings of Case 4 showing increasing degrees of distal bulging with greater suprapubic pressure.

Case 5. T.H. was born after an uneventful full-term pregnancy and a normal vaginal delivery. He presented at six months of age with fever and irritability secondary to urine infection. An US showed bilateral hydrenephrosis, a thick walled bladder and a dilated urethra. His renal function was normal. A suprapubic catheter was inserted followed by an MCU (Fig.9A). Obstruction confirmed, an endoscopy was performed. After viewing the membrane (Fig.9B), an 8FG catheter was passed, which split the obstruction into two "leaflets" (Fig.9C). Because of the small size of the phallus the Whitaker diathermy hook was used to further disrupt the membrane; the "leaflets" become more obvious and the sphincter could be seen to come to rest part way along these folds.
Figure 9A: The cystogram of Case 5: again the typical features are seen similar to those in Fig. 7 and 8.

Figure 9B+C: The endoscopic findings of Case 5, before (B) and after (C) the membrane has been converted from a membrane to "valve leaflets" by the passage of an 8FG catheter.
Case 6. O.F. was born at term from a normal pregnancy and delivery, including a normal US at 17 weeks gestation. He presented at three months of age, but first became unwell when 10 days old. His episode of sepsis, at that time, was attributed to Staphylococcus Aureus enterocolitis. Two urinary tract infections ensued three and seven weeks later. Ultrasound then showed bilateral hydronephrosis and a thick walled bladder. An MCU at the referring hospital demonstrated an abnormal bladder and posterior urethra. Unfortunately, no films were taken with the urethral catheter removed. Endoscopy was undertaken without further imaging or urethral catheter insertion. A membrane was seen attached to the verumontanum with a slightly larger defect than seen in the other index cases. The obstruction was resected with the infant resectoscope.

Case 7. J.H. presented at the age of six weeks following a grand-mal convulsion secondary to renal failure and hyponatraemia. His bladder was drained for two days with a fine feeding tube, and his Cr decreased from 270 mmol/l to 107 mmol/l. The patient was then transferred, but not immediately to urological care. An US showed bilateral hydronephrosis, hydroureters, a thick walled bladder and a dilated posterior urethra. An MCU was performed through a urethral catheter and cystourethroscopy was undertaken later the same day. An obstructing membrane, with a pinhole posterior orifice was larger than in those boys who had not had preoperative urethral catheterization; an 8FG catheter partly split the membrane anteriorly. The Whitaker diathermy hook was used to further disrupt the membrane which produced the two "valve leaflet" appearance that most interpret as a "Type I valve". The sphincter was then visible lying on the "leaflets", between of the verumontanum and the original membrane (as in Cases 2 and 3).
Retrospective Study

Before the introduction of the suprapubic drainage without urethral instrumentation, 12 earlier patients had endoscopy at the time of a Whitaker hook procedure. This latter information was collected retrospectively, and is therefore, less noteworthy than the cases presented in detail above, but it does support the view that the obstruction of the posterior urethra is altered by the presence of an indwelling urethral catheter, or the passage of a urethral instrument as seen in the cases above. All 12 boys had a urethral catheter for a contrast MCU and 11 had urine drainage via a urethral catheter to allow stabilization prior to their operation. Nine of the 12 were labelled as "Type I valves", but all had a urethral catheter in situ for a number of days, whereas two other boys with minimal manipulation of their urethra had an almost complete membrane, similar to those boys studied prospectively. The last of the 12 had a membrane with a hole (called a "Type III obstruction" by the surgeon) converted to the appearance of valve leaflets (called a "Type I obstruction" by the surgeon) by the passage of the Whitaker hook. Preoperatively, this boy had only one day of urethral catheter urine drainage.
CONGENITAL POSTERIOR URETHRAL OBSTRUCTION
Adelaide Children's Hospital Patients

Introduction

The Adelaide patients with a posterior urethral membrane, presented with symptoms and signs of outflow obstruction or were found to have an incidental abnormality on endoscopy; the development of a prospective database and video recording of endoscopies facilitated these observations. The conclusions reached were enhanced by the involvement in the prenatal diagnosis and therapy group, in the recently amalgamated Women's and Children's Hospital. The use of pre-emptive perineal US and an approach of minimal intervention in the urethra before endoscopic video, further amplified the quality of documentation.

A review of the literature had been conducted prior to the first reporting of the observations in Adelaide, a careful study of which indicated that the classification of congenital posterior urethral obstruction should be revised in line with the improved technology for investigating, recording and treating the abnormality. Thus the term Congenital Obstructive Posterior Urethral Membrane (COPUM) was coined, which most specifically addressed the finding of the conversion of a posterior urethral membrane to valve leaflets. The term COPUM will usually be used in place of "valves" for the remainder of the text.
Materials and Methods

Between October 1990 and October 1995, 22 boys were diagnosed and treated for congenital obstruction of their posterior urethra; a further 14 were thought to have a possible obstructive change on their cystogram and six were included on the basis of endoscopic findings alone. Those with a prenatal diagnosis, who did not survive were not included. All boys had a video recording of the first endoscopic entry into the urethra, after usually minimal or no preoperative instrumentation. Because of the different findings in the three subgroups, the patients will be presented in their groups.

During review of the video, the position of the proximal extent of the external sphincter, the nature of the verumontanum, the appearance of the folds from its distal extent and the position of the posterior urethral membrane were noted and subsequently entered onto a database. Detailed documentation of the membrane included its reaction to suprapubic pressure, the size of the defect at its posterior edge, the dilatation of the posterior urethra and any additional features.

Pre-operative cystograms were performed and reviewed at the time of further study of the endoscopy in all 42 boys.
Results

The results for the assessment of the posterior urethral membranes will be presented according to the three arbitrary groupings, based on the endoscopic appearance. The demographic features of each group and the detailed case descriptions of those with an obstructive lesion are given. The relationship between the external sphincter and the membrane is presented on page 80 as a compilation of all 42 cases.

Obstructive Membranes

Clinical Findings

This group of 22 cases presented at a median age of 3.7 months, with a range from 0.3 to 171.4 months, with the presenting problems as shown in Table 1.

<table>
<thead>
<tr>
<th>Condition</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Urinary Tract Infection</td>
<td>11</td>
</tr>
<tr>
<td>Prenatal Hydronephrosis</td>
<td>8</td>
</tr>
<tr>
<td>Incontinence</td>
<td>1</td>
</tr>
<tr>
<td>Other</td>
<td>2</td>
</tr>
</tbody>
</table>

Table 1: The presentation of those boys who were found to have an obstructive posterior urethral membrane.

The following is a summary of the cystogram and endoscopic findings for the 22 boys with an obstructive anomaly.

Cystogram Findings

All these boys had a pre-operative cystogram. The visibility of the verumontanum and the distal folds is given in Table 2. The proximal extent of the external sphincter was definitely identified in 16, probably identified in two and not seen in four (see the case reports for illustrations).
Table 2: The correlation of the findings for the verumontanum and the folds running onto the COPUM for the 22 with an obstructive lesion.

The degree of dilatation of the posterior urethra ranged from minimal in five to gross dilatation in two; seven boys had a cystogram which was "text book". Illustration of the variation is given in the figures for the case reports.

**Cystoscopy Findings**

Five of the cases had a Cobb's collar, in addition to the COPUM, four with a moderate narrowing and one minimal. The verumontanum was flat in two and two had a small cyst on the apex.

The external sphincter was seen in 17, all above the obstruction. The sphincter was seen to come down onto the folds running from the verumontanum in four cases. All boys had a membrane with a posterior hole and a number converted to the appearance of valve leaflets after passage of the cystoscope through the defect. Illustration of these findings are given in the following case reports.
The clinical details of the 22 cases of obstructive membranes are as follows;

**Case 1:** BW presented with a urinary tract infection at two months of age. On US examination he was found to have a minimally thickened bladder wall and left hydroureteronephrosis. A cystogram identified left VUR and obstruction of the posterior urethra (Fig.10) which, on cystoscopy, was seen to be a membrane with a posterior hole just below the verumontanum. The obstruction was resected endoscopically and the urethra was satisfactory on repeat cystogram and endoscopy three months later, when his GFR was 125 mls/min/1.73m² and his kidneys functioned equally.

![Figure 10: The cystogram for Case 1, who was found to have a pinhole meatus at the back of the obstructing membrane on cystoscopy.](image)

**Case 2:** At 5 weeks of age JB developed a vomiting illness and became dehydrated. On admission elsewhere he was found to be hyponatremic and had echogenic, hydronephrotic kidneys on US. A nuclear medicine scan showed bilateral poor function and dilated ureters. He was transferred for further management, when a perineal US showed a dilated
posterior urethra and obstructing membrane similar to Fig. 11. A cystogram confirmed the
diagnosis of posterior urethral obstruction and endoscopy verified the membranous nature
of the obstruction which was then fulgurated. On follow-up at 6 months of age, his GFR
was 81 mls/min/1.73m², his urethral anatomy more normal and his hydroureteronephrosis
improved.

Case 3: BD presented at nine months of age with a febrile convulsion secondary to a
urinary tract infection. Bilateral hydroureteronephrosis and a thick walled bladder were
seen on abdominal US, and a dilated posterior urethra and obstructing membrane were seen
via a perineal US (Fig. 11A) and confirmed with a cystogram. The obstructing membrane,
demonstrated endoscopically, was below the verumontanum, had a pin-hole meatus at the
posterior edge and split on passage of a 10FG cystoscope (Fig. 11B). The membrane was
fulgurated, and subsequent renal function has been satisfactory, after an initial post
obstructive diuresis.

Figure 11A: A perineal US of Case 3 showing the obstructing membrane at the lower end of the
dilated posterior urethra.

Figure 11B: The verumontanum can be seen with rough edges from splitting in the membrane.
Case 4: GK, a boy with trisomy 21, developed urinary retention at six weeks of age. Ultrasound of his perineum identified a dilated posterior urethra (similar to Fig.11), leading to the insertion of a suprapubic catheter. A cystogram and endoscopy documented a posterior urethral obstruction due to a membrane below the level of the veru. His initial recovery was delayed by a degree of vesicoureteric obstruction, which settled, and he was discharged with a Cr of 70 mmol/l two weeks after the disruption of the obstructing membrane. He remains well with satisfactory bladder and kidney function.

Case 5: JH, first born of a normal term, non-identical twin pregnancy, was thought to have unilateral hydronephrosis and normal amniotic fluid on two late gestation prenatal scans. On day five of life he became febrile and *Staph. aureus* was grown from his urine. Subsequent US noted bilaterally echogenic renal parenchyma, a moderately dilated left renal pelvis and ureter, a grossly thickened bladder wall and a transiently dilated posterior urethra. A cystogram was attempted, but a catheter could not be passed into the infant’s bladder. Therefore, a retrograde cystourethrogram was performed which showed an obstruction just below the verumontanum, plus a distal membranous structure extending down to the bulbar urethra (Fig. 12A). Endoscopy revealed the distal part of the abnormal anatomy to be a diverticulum of the posterior urethral wall, below the obstructing membrane. The diverticulum, which had a proximal hole in its anterosuperior surface and a smooth edged distal defect, did not appear to be obstructive. The diverticulum was connected to the obstructing membrane, from below and anteriorly, by two paramedian reinforcements (Fig.12B) in a similar fashion to the paramedian ridges which ran down and forward from the verumontanum (Fig.13 - Case 6). The obstructing membrane (Fig.12C) and the position of the external sphincter were otherwise as described for the other boys in this group. The obstructing membrane was fulgurated and a nuclear scan three days post-operatively showed no function on the right kidney and poor function on the left. His Cr stabilized at 125 mmol/l.
Figure 12A: Case 5. The obstructing membrane and the distal diverticulum are seen on the cystourethrogram.

Figure 12B: The line drawing shows the distal diverticulum, attached to the inferior surface of the obstruction. The fenestrations represent the caudal and cranial openings in the anterior wall of the diverticulum. The paramedian reinforcements of the membrane run down from the verumontanum.
Figure 12C+D: Case 5. The distal extent of the diverticulum (C) which ran between the COPUM and the bulbar urethra. The two halves of the anterior wall of the diverticulum can be seen to meet at the mid-point of its length [lower edge of picture] (D).

Figure 12E: Case 5. The COPUM is seen with the leaves of the distal diverticulum running onto the under surface.
Case 6: CW was irritable for two weeks before he presented as a sick, four month old infant. He had normal kidneys on US, at 18 weeks gestation and was thought to have a normal urinary stream postnatally. On investigation he was found to have an Enterococcal urinary infection, bilateral hydroureteronephrosis and a large bladder. An antegrade cystogram, through a suprapubic catheter, showed a typical congenital obstructing posterior urethral membrane (Fig.13A). When biochemically stable, his urethra was examined endoscopically, and the obstructing membrane was seen to have a small posterior defect (Fig.13), to prolapse as suprapubic bladder compression was applied (Fig.13C) and to changed to the "valve leaflets" appearance on passage of the cystoscope (Fig.13D). The obstruction was fulgurated and he is currently undergoing further bladder and upper urinary tract investigations and management, which has included a ureterocystoplasty.

Figure 13A: An antegrade cystogram through a suprapubic catheter, showing the typical features of congenital obstruction of the posterior urethra and vesicoureteric reflux.
Figure 13B+C: B. The non-ballooning appearance, with a 4FG ureteric catheter passed through the posterior defect in Case 6. C. Distal ballooning was seen with suprapubic pressure.

Figure 13D: The proximal end of the external sphincter was proximal to the obstructing urethral membrane, as seen here for Case 6. The paramedian membrane reinforcements can be seen sweeping down, through the sphincter, from the verumontanum.
Case 7: AB was found to have bilateral hydronephrosis and a thick walled bladder at 24 weeks gestation. Right renal aspiration at 28 weeks recorded normal fetal renal biochemistry. As the amniotic fluid remained stable the pregnancy was continued to 37 weeks without further intervention. Postnatal investigation confirmed gross right uretero-hydronephrosis and echogenic parenchyma and minimal renal tissue on the left. He was further investigated with an antegrade cystogram (Fig.14A) and urethrocystoscopy. An obstruction at the level of the bulbar urethra, seen on the cystogram, was noted to consist of a membrane sweeping down from the level of the verumotanum with a 3mm defect present in its posterior aspect. A smooth-edged, rounded defect, in the thin anterior portion of the membrane, was also seen (Fig.14B); the superior surface of the membrane was seen after passing the cystoscope through that anterior defect, and again, the posterior defect was visualized (Fig.14C). Much of the membrane was disrupted by passage of the endoscope, and the remainder was resected with diathermy. This boy is continuing to undergo further renal functional assessment after an early stabilization of his Cr at 70 mmol/l; he is one of the three COPUM boys who has had a ureterocystoplasty.

Figure 14A: The distal obstructing membrane in the antegrade cystogram of Case 7, swept down from the verumotanum and had the two defects seen in Figures 14B,C+D.
Figure 14B,C+D: The "fixed balloon" of Case 7 has a defect in the thin anterior portion (B) and the typical posterior hole seen from proximal (C) and distal (D) to the obstruction.
Case 8: AC was found to have hypertension, microscopic haematuria, a right duplex kidney with lower pole reflux and huge distension of his posterior urethra (Fig.15) after he presented with lower abdominal pain. He was managed initially by incision of his COPUM which had the muscular appearance as seen with JL (Case 16). He subsequently had a check cystoscopy, lower pole ureteric reimplant and left upper pole heminephrectomy. He was well, with good bladder control and stable renal function, two years after initial presentation.

Figure 15: A cystogram which shows huge distension of the posterior urethra (A). The cystoscopy showed a more crenated appearance to the obstructing lesion (B).
Case 9: AD was transferred for care at 11 days of age with pyelonephritis and thrombocytopenia. Investigation identified a single right, echogenic, hydrenephrotic kidney and urethral obstruction (Fig. 16). An endoscopy confirmed a COPUM, which had the appearance of a membrane with a posterior hole, changing to "valve leaflets", on splitting of the membrane (Fig.17). The obstruction was treated by incision at the five, seven and 12 o'clock positions, followed by a check cystoscopy and circumcision three months later. He was last reviewed at 2.5 years of age, at which time his GFR, renal US and bladder function appeared normal.

Figure 16: The neonatal cystogram shows a marked calibre change, but less significant posterior urethral dilatation than typically seen.
Figure 17A: The posterior defect in the COPUM, before instrumentation in Case 9.

Figure 17B: The sphincter can be seen adjacent to the folds from the verumontanum; the "membrane with a posterior hole" appearance (above) has been changed to "valve leaflets" by a change of position of the cystoscope.
Case 10: JD was found to have hydronephrosis early in pregnancy with significant progress of the parenchymal thinning and hydronephrosis, resulting in the early delivery of the child at 35 weeks gestation. A cystogram confirmed the presence of a urethral membrane, without marked dilatation of the posterior urethra, but with high grade VUR (Fig.18). The serum creatinine remained normal during his early treatment with a 5FG feeding tube for urethral drainage. The initial treatment was a Fogarty balloon disruption of the obstruction, followed by improvement in the dilatation of the upper tracts and a cystoscopy to fulgurate remnants of the membrane, which had a typical, but less obstructive appearance (Fig.19). Bilateral ureteric reimplants have subsequently been performed for VUR which had not resolved with the normalization of his urethra. His GFR was 77 ml/min/1.73m² and his renogram showed minimal parenchymal loss.

Figure 18: The neonatal cystogram in this boy showed minimal dilatation, but a definite membrane.

Figure 19: The endoscopic appearance after Fogarty balloon disruption confirmed the presence of a COPUM as the cause of the bilateral VUR.
Case 11: TH was referred from a peripheral hospital with failure to thrive and a urinary tract infection (UTI). His cystogram showed an obstructing lesion in the posterior urethra, with minimal dilatation proximally and no vesicoureteric reflux. Despite the minimal changes on the cystogram (Fig.20), a typical COPUM was identified endoscopically (Fig.21). The lesion was difficult to fulgurate because of access problems, therefore the urethra was also dilated. The follow-up cystoscopy was not performed as the patient moved interstate from where he was reported to be doing well, with good bladder and kidney function.

Figure 20: The cystogram of Case 11 shows minimal dilatation, a membrane at the level of obstruction shown on the cystoscopy (Fig.21) and a more distal Cobb's collar.

Figure 21: The typical membrane appearance found in Case 11.
Case 12: MH presented with fever, a urinary tract infection and poor stream at the age of 12 years. He had been born premature, but, apart from asthma, had remained well since the neonatal period. A cystogram delineated the posterior urethral anomaly poorly, because of the dilute contrast used. However, he was shown to have a posterior urethral obstruction, a large posterior urethral diverticulum and bilateral VUR. At endoscopy, a membrane with a posterior hole (Fig.22) was identified, but initial fulguration failed to improve his bladder outflow and further endoscopy and fulguration were required on two occasions. Transvesical removal of the infected posterior diverticulum was also performed and in addition, bilateral ureteric reimplants were necessary. He has subsequently been well with good renal function and improving bladder function.

Figure 22: A posterior defect is seen to be partly inflamed in a boy who presented with a urinary tract infection (A). The relative dimension of the membrane and the defect in the posterior lip is indicated by the 4FG ureteric catheter (B).
Case 13: WJ presented at the age of 10 days with a UTI and was investigated with an US and MCU. A membrane was seen in the posterior urethra, but without marked dilatation of the posterior urethra (Fig.23). On endoscopy, a definite, but less obstructive COPUM was seen (Fig.24). Fulguration produced a satisfactory result endoscopically, normal kidneys on US and good renal function.

Figure 23: A cystogram at six weeks of age, showing little dilatation of the posterior urethra, but a membrane of the COPUM.

Figure 24: The endoscopic view of the obstruction in Case 13, (A) before passage of the cystoscope through the posterior defect and (B) after instrumentation.
Case 14: MK, one of twins, was found to have prenatal hydronephrosis which appeared to be progressing in utero. Fortunately for the twin with urethral obstruction, the pregnancy delivered spontaneously at 29 weeks gestation, without significant complications of prematurity. The small size of this boy, and the presence of a minute urethral meatus associated with a minor hypospadias, necessitated the formation of a vesicostomy. He subsequently underwent a cystogram (Fig.25) via the stoma, and urethral endoscopy fulguration (Fig.26), both of which demonstrated the typical features of a COPUM.

Figure 25: The cystogram, via the vesicostomy, in Case 14, showing the usual change of calibre with posterior urethral obstruction.

Figure 26: The endoscopic appearance of Case 14 shows contraction of the proximal end of the external sphincter onto the crista urethralis, just below the verumontanum.
Case 15: DK, also one of a set of twins, presented with a prenatal diagnosis of hydronephrosis. The twins were delivered by caesarian section at 38 weeks gestation, following which an US, MCU and renal function studies were performed. His Cr initially increased to 175 mmol/l and thereafter was normal. His cystogram showed the typical changes of posterior urethral obstruction (Fig.27) and his cystoscopy showed a membrane with a hole, the sphincter above the obstruction and the conversion of the lesion to typical "valve leaflets" by the passage of the scope (Fig.28).

Vesicoureteric reflux was present on the pre-operative cystogram, but not on the follow-up studies and his urethra was satisfactory at the time of re-endoscopy. He remains well and thriving, with a normal GFR and bladder dynamic function, four years after his initial presentation.

Figure 27: The classic cystogram of congenital posterior urethral obstruction from Case 15.
Figure 28A: Case 15 - The level of the obstruction on the endoscopy, showing a membrane with a hole at the posterior edge.

Figure 28B: Case 15 - alteration of the position of the scope gives the appearance of "valve leaflets" and the sphincter can be seen above the level of the obstruction.
Case 16: JL had previously had an endoscopy 12 months before he presented to me with what must have been minimal previous treatment of a posterior urethral obstructing lesion. He originally presented at the age of 14 years with a urinary tract infection and septicaemia. Endoscopy identified an obstructing membrane in his posterior urethra, with a different appearance to the majority of the others and similar to Case 8 - as shown in Figure 15. The region of obstruction appeared crenated, as if to contain muscular elements. His stream was significantly improved by the intervention which consisted of fulguration of the membrane and insertion of a urethral catheter for three days. Check cystoscopy, three months later, showed a good endoscopic result. Follow-up studies show the right kidney to have reduced but stable function, his GFR is normal at 123 ml/min/1.73m² and he continues to void normally.

Case 17: AL, a two year old boy, was prenatally noted to have an abnormal bladder and upper renal tract. Postnatally, he was found to have high grade VUR with urethral atresia and a large, trabeculated, thick-walled bladder. He had ureteric reimplants and a vesicostomy at four months, as initially it was felt that bladder augmentation would not be required. Subsequently, his urethra was identified to have a COPUM proximal to his urethral atresia, for which he was treated. His vesicostomy was closed at 18 months and he was commenced on intermittent catheterization. He was not initially identified to have a COPUM because of the poor filling of the posterior urethra during the neonatal cystogram. His upper renal tract continued to dilate, despite the cessation of the reflux and his bladder had become thick walled, small volume and high pressure, for which he has had a successful ureterocystoplasty.
Case 18: RM, presented to my care in 1992, at the age of seven years, at which time he was suffering from incontinence and had a poor stream. His symptoms had been constant all his life. He initially presented for medical care after a prenatal diagnosis of hydronephrosis and subsequently had a cystogram and a diagnosis of posterior urethral obstruction. The details of the endoscopic findings at that time were not specifically noted. As a neonate he had a transient rise in his Cr after which his Cr has always been normal and his GFR at nine years of age was 121 ml/min/1.73m².

A cystogram performed as a follow-up to his original intervention continued to show a marked change in calibre, however, no action was taken. A repeat MCU indicated that the obstructive changes were still present (Fig.29), therefore, a video recorded endoscopy was undertaken, identifying a COPUM, with both a "membrane with a posterior hole" (Fig.30A) and "valve leaflets" (Fig.30B) images recorded. Also, the proximal portion of the external sphincter was above the level of the obstruction (Fig.30C). His incontinence, the posterior urethral dilatation on MCU and the membrane obstruction on endoscopy were all resolved by the further intervention.

Figure 29: Case 18 - A cystogram with the typical features of posterior urethral obstruction, despite the previous intervention.
Figure 30: Case 18 - A view of the membrane from below (A) and part way through the posterior defect (B), giving the impression of a "membrane with a posterior hole" and "valve leaflets" in the one patient.

Figure 30C: The proximal portion of external urethral sphincter sits adjacent to the folds running from the verumontanum to the proximal surface of the COPUM.
Case 19: JS, was initially managed in another centre, supposedly with fulguration of a posterior urethral obstruction. He had presented with abdominal pain, fever and dysuria at the 4.5 years of age. Following the initial fulguration, he became incontinent. An MCU showed that the obstruction had not changed significantly by the previous intervention (Fig.31). The boys renal function was normal and his bladder was of good volume and minimally trabeculated. Endoscopy also suggested that the membrane had not been significantly altered by the previous intervention (Fig.32); fulguration achieved an improvement in the endoscopic and MCU appearance of his urethra and his incontinence was resolved.

Figure 31. Case 19 - MCU showing the persistence of obstruction at the level of the membrane.

Figure 32. Case 19 - endoscopy shows the membrane obstruction, with the typical posterior hole only minimally altered.
Case 20: CP, was noted to have marked progression of hydronephrosis on prenatal US, with a dilated bladder and upper urethra. The US at 32 weeks gestation showed increased echogenicity and dilatation, therefore early delivery was arranged. The patient initially had mild respiratory distress and was managed urologically with the insertion of an 8FG feeding tube. On day three, his Cr was elevated to 190 mmol/l, then settled and stayed normal. His respiratory status rapidly improved and he went on to have a cystogram which diagnosed the urethral obstruction and the presence of grade V, bilateral vesicoureteric reflux. A Fogarty balloon disruption of the COPUM was performed and subsequently a cystoscopy and fulguration of the remnants was carried out. The ablation of the obstruction failed to resolution the VUR, therefore he had bilateral ureteric reimplants at four months of age. He remains well and thriving with a GFR of 115 ml/min/1.73m² at two years of age.

![Image](image-url)

Figure 33; The prenatal US showing a dilated bladder (A) and echogenic kidneys (B).
Case 21: BM, was diagnosed prenatally to have posterior urethral obstruction. Marked progression of the prenatal hydronephrosis (Fig. 34), urethral dilatation (Fig. 35) and increasing echogenicity of both kidneys prompted bladder aspiration with a view to early delivery. The bladder tap showed good quality urine with a low sodium and β2-microglobulin, therefore the baby was delivered at 32 weeks gestation. Mild lung disease and the size of the genitalia precluded early endoscopic intervention, necessitated management of the infant with urethral catheterization (Fig. 36). The subsequent cystogram showed the urethral membrane and bilateral high grade VUR. Because of the minimal membrane on the urethrogram and the marked improvement in the renal function and US abnormality of the kidneys, endoscopic intervention was deferred until the age of three months. A significant residual membrane was fulgurated, bilateral ureteric reimplants performed for persistent VUR and circumcision for phimosis were subsequently performed. His GFR improved from 30 ml/min/1.73m² at the age of two months to 52 ml/min/1.73m² at one year.

Figure 34: Bilateral hydronephrosis, increased echogenicity and a dilated bladder seen at 20 weeks gestation in Case 21.
Figure 35: A prenatal view of the bladder shows the dilated bladder and urethra.

Figure 36: The thick walled bladder seen postnatally while the bladder was on drainage.
Case 22: HM, was delivered at 36 weeks gestation because of progressive hydroureteronephrosis and a dilated bladder, which was shown on the post delivery cystogram to be due to posterior urethral obstruction (Fig. 37). A Fogarty balloon disruption of the lesion was performed and the remnants of the urethral membrane (Fig 38) were resected one month later. Three months later he had a circumcision and a further partial resection of the membrane. He remains well with a normal GFR and Cr at 12 months of age.

Figure 37: Cystogram showing less dramatic urethral calibre change at the level of the obstruction, than the classic appearance.

Figure 38: Cystoscopy showing a membrane with the posterior hole in the posterior urethra.
Moderate Membranes

Clinical Findings

The 14 boys who were found to have a moderate membrane were first endoscoped at a median age of 52.8 months, with a range of 1.7 - 126.8 months. The nature of their presentation is given in Table 3.

<table>
<thead>
<tr>
<th>Presenting Complaint</th>
<th>No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Urinary Tract Infection</td>
<td>8</td>
</tr>
<tr>
<td>Prenatal hydronephrosis</td>
<td>2</td>
</tr>
<tr>
<td>Incontinence</td>
<td>2</td>
</tr>
<tr>
<td>Abdominal pain + dysuria</td>
<td>2</td>
</tr>
</tbody>
</table>

Table 3: The presenting complaint for the 14 boys with a moderate membrane.

Cystogram Findings

Of the 12 that had a pre-operative cystogram, the verumontanum was prominent in six and moderately well seen in the remaining six. There was no evidence of the moderate COPUM in four cases of the 12 cystograms, five had a definite membrane (Fig.39) and three only had a minimal impression of the existence of the posterior urethral membrane. Folds running from the verumontanum were only seen in three cases and only in one of these were they at all prominent (Fig.40).

The proximal extent of the external sphincter was seen in nine of the 12 cystograms; a minimal indentation in four and a marked indentation in five (Fig.41). Where both the
sphincter and the COPUM were seen, the former was proximal to the latter.

In addition, seven of the nine with a cystogram and an endoscopically identified Cobb's collar had some evidence on the cystogram.

Figure 39: A moderate membrane visible on the preoperative cystogram.

Figure 40: Folds running from the verumontanum to the membrane can be seen.

Figure 41: One of the five cases with a moderate COPUM in which the proximal extent of the external sphincter can be easily visualised on the cystogram.
Cystoscopy Findings

The penile urethra was normal in all but three of this group; one had a minor cyst, one a syringocele and the other, minimal urethritis.

A Cobb's collar was seen in 11 boys, either muscular (9) or of uncertain nature (2) and all were either a minimal or moderate narrowing, none of which required treatment.

The verumontanum was of the normal configuration in the majority, as a mound in the posterior urethra. In four boys the verumontanum was relatively flat, two of which were associated with a long crista running distally from the verumontanum before diverging to form the folds on the proximal surface of the COPUM. The sphincter was visualised close to the COPUM in four cases, with the anterior aspect of the membrane appearing to have some crenation, as if to have some muscle in the anterior aspect of the membrane in those boys. The remaining 10 membranes were seen distal and separate from the proximal extent of the external sphincter. Examples of the endoscopic view are shown in Fig.42 + 43.

Figure 42: The close up view of a moderate COPUM (A), which can be seen from a more distal view in B.
Figure 43A+B: Two further cases of moderate COPUM. (A) shows the proximal end of the external sphincter in contact with the folds coming from the distal end of the verumontanum.
Minimal membranes

Clinical Findings

All six boys who were found to have a minimal membrane presented for investigation because of other pathology, with the urethral anomaly as an entirely incidental finding. The boys presented at a mean age of 50.7 months, with a range of 12.1 to 146.4 months, with either a urinary tract infection, prenatal hydronephrosis, day-time incontinence, haematuria or for the investigation of hypospadias repair complications. A cystogram was performed in four boys, two of whom had some indication of the presence of the COPUM (Fig.44). The proximal end of the external sphincter was easily identified in three (Fig.45).

Endoscopically, the verumontanum was normal in all six boys and the proximal end of the external sphincter was easily identified above, and separate, to the membrane elements (Fig.46 + 47). Each of these boys had more prominent membrane elements than those of the 119 normal endoscopies who were found to have prominent folds running distal from the verumontanum.

A muscular Cobb’s collar was seen in two boys; minimal in one and moderate in the other. Neither the COPUM nor the Cobb’s collar in any of the six boys warranted any intervention.
Figure 44: A cystogram showing the position of the minimal membrane, distal to the indentation caused by the external sphincter.

Figure 45: The proximal end of the external sphincter is shown occluding the urine stream, just above a minimal membrane.
Figure 46: A minimal fold can be seen extending from the distal end of the crista urethralis, through the urethral sphincter. The bladder is empty in this view and filled in the figure below.

Figure 47: The thin rim of membrane (shown in Fig. 46) when the bladder is maximally filled. The sphincter is just proximal and the folds from the verumontanum have been flattened out by the fluid pressure.
Position of the External Sphincter

Cystoscopic Findings

In the forty-two boys who had a membranous lesion in the posterior urethra, 22 were considered obstructive, 14 were less significant, but possibly obstructive and six were minor incidental membranes. In 36 of the cases, the proximal extent of the external sphincter was identified proximal to the posterior urethral membrane (Fig. 48, 49 + 50). In six cases, there was a limited view through the defect in the lesion and the proximal extent of the external sphincter was possibly, but not reliably seen, however, the characteristic external sphincter was not present distal to the membrane in these cases.

Figure 48: The proximal extent of the external urethral sphincter is seen close to the folds which run from the verumontanum to the membrane (A). The sphincter on the folds, when the water flow is turned off, is shown in B.
Figure 49: Another obstructed urethra case in which the proximal extent of the external urethral sphincter is seen adjacent to the folds coming from the verumontanum.

Figure 50: A non-obstructive membrane with the sphincter well shown, proximal to the membrane.
Radiological Findings

Of the 38 cystograms, four were of poor quality because of inadequate filling of the posterior urethra. In these three, neither the level of obstruction, nor the location of the external sphincter could be seen. The level of the membrane was evident on the remaining 35 studies, and evident in all those with obstruction.

In thirty-five cases, the filling defect of the verumontanum was demonstrated to be located above the level of the membrane, as was the proximal part of the external sphincter in 31. An abrupt change in calibre was seen in those with marked dilatation of the posterior urethra secondary to the obstruction (Fig.51) and a more tapering change was found in those with lesser effects of the obstruction (Fig.52).

Eighteen cystograms showed linear filling defects extending inferiorly from the distal extent of the verumontanum to the proximal surface of the posterior urethral membrane, correlating with the cystoscopic finding of folds attached from the verumontanum to the membrane.

Figure 51A: An MCU of a markedly dilated urethra, before sphincter contraction.

Figure 51B: An MCU of a markedly dilated urethra, after sphincter contraction.
Figure 52A: An MCU of a minimally dilated urethra, before sphincter contraction.

Figure 52B: An MCU of a minimally dilated urethra, after sphincter contraction, showing the marked change of calibre at the site of the contraction.
DISCUSSION

Classification

The first attempt to classify congenital posterior urethral obstruction was by Tolmatschew in 1870 [40]. Further refinement was offered by Young et al. in two papers in 1919 [1] and 1929 [2] with a three part classification based on the initial 12 patients and a review of the literature. They further subdivided the Type I and III groups; Type III lesions were thought to consist of a centrally perforated membrane either above (Type IIIa) or below (Type IIIb) the verumontanum, without attachment to the verumontanum (Fig.53).

Figure 53: Young's classification from 1919; note the six part classification from 12 patients.

A total of 21 cases were reported in the two papers (15 "Type I", two "Type II", and four "Type III"). Eight were cystoscoped (two of these also had an autopsy), eight were diagnosed by the antegrade or retrograde passage of a finger or the urethral "Punch" (one had a late autopsy). The remaining five patients had an autopsy only. None of the four patients with a "Type III" valve were diagnosed by endoscopy, and only one was diagnosed
only one was diagnosed at autopsy, i.e. three did not have any visualisation of the lesion.

Of the eight patients cystoscoped, five had prior urethral manipulation and, from the operative reports, the remaining three had the cystoscope passed into the bladder before viewing of the urethra during withdrawal of the instrument. Referring back to the 1919 paper, only three of the 12 cases had been cystoscoped, and these patients were 17, 26 and 42 years old, one with previous venereal infection. Five were described as Type I, one Type II, three were Type III and three were not classified. Young et al. presented an extensive literature review in 1919 which, unfortunately, does not indicate which of the cases reviewed were assigned to each part of the classification, thus precluding an accurate critical analysis. Critique is also hampered by only 20 of the listed cases being included in the classification table, and the number of cases in the table and the text differ; notably, an 85 year old patient was included in the discussion. Young made two comments of particular note in his original paper on this subject which indicate the need for a change in perspective with improved technology [1]: "cystoscopy furnishes little information regarding the type of urethral anomaly", which most would now find rather contentious, and "little or no difficulty is encountered in passing a catheter, but the ease of instrumentation is by no means an index of the degree of obstruction". The latter statement has been used by others to imply support for the obstruction being valvular, whereas the current evidence would suggest that it is either a flimsy membrane or an elastic membrane orifice which allows for the easy passage of the catheter.

It was from Young's work that the term Posterior Urethral Valves was introduced, however, the technique of dissection, the prior urethral instrumentation and the technique of endoscopy mitigated against complete elucidation of the anatomy. In the light of the findings in this thesis, these factors would have had a significant effect on interpretation of the anatomy, as outlined below.

The specimens, as illustrated in Young's papers, show that the urethra was open by a
midline anterior approach, leaving the posterior wall intact, with the potential for a membrane obstruction to be split and give the impression of a valvular obstruction. A number of other early papers also showed such autopsy specimens or illustrations, including those by Wilckens in 1910 [52], Heinecke in 1913 [57], Hinman and Kurtzmann in 1925 [36], Kretschmer and Pierson in 1929 [61], Campbell in 1931 [71], Stephens in 1963 [18] and Williams in 1968 [30]. The concept of splitting a membranous obstruction between two paramedian reinforcements is supported by the two diagrams given in Lederer's 1911 paper: a view from above shows a membranous obstruction and later in the dissection (after an anterior midline incision), an anterior view shows a split membrane with two "valve leaflets" [55].

Type II lesions (obstruction above the level of the verumontanum) were, generally, only recorded in the early reports [4,70,71,124,125] and, despite the recording by other than Young et al., many authors have since suggested this lesion to be a misinterpretation of the secondary effects of the more distal obstruction [11,126-128]. Stephens [18] did not find any Type II valves in his series and concluded that it was "possible that a false endoscopic diagnosis in the absence of radiographic confirmation led to the erroneous description of the so-called Type 2 (sic) valve of Young et al. (1919)". I did not observe any case with a Type II lesion in the 210 male patients cystoscoped during the five year period studied.

Field and Stephens [129] later provided evidence of a prolapsing form of the Type III lesion. This appearance was seen in AB (COPUM case 7, Fig.14), who had a fixed membrane with attachments to the verumontanum and, like one of the six cases presented by Stephens [18,129], he had two orifices in the membrane. The central hole in the Adelaide Case 7, appeared to have been a thin membrane which had worn through; the original defect being a posterior hole in a typical, but prolapsed COPUM. The additional finding of significant, transient membrane ballooning in response to suprapubic pressure in other cases seems consistent with the ability of the COPUM to prolapse down the urethra (see Fig. 8C, 8D and 13C). The differentiation between the more distally located lesion
and a COPUM which has prolapsed, is the connection of the COPUM to the verumontanum via folds. It seems that Stephens grouped together lesions with and without a connection to the verumontanum. The strength of his conclusion is weakened for two reasons: firstly, six of his 10 cases did not have a cystogram (they had an autopsy only); and secondly, four were found to have a narrowing at the junction of the membranous and bulbar urethra on a cystogram, but were not endoscoped and survived [18]. These latter boys were treated successfully by dilatation, however, the lack of the combination of cystogram and cystoscopy impacts on the interpretation of the data.

A different posterior urethral dissection method, which produced an appearance of a diaphragm, was first used by Jarjavay in 1856 [39]. The diagram by Lederer in 1911 was the next such publication [55], but it was not until the post-mortem studies of Presman in 1961 [3] and Robertson and Hayes in 1969 [5], that it was suggested that Young's classification is incorrect. Prior to that, however, there were a number of studies which reported the presence of Type III lesions [93,127,130], although, a very small proportion of the cases were presented and many reports discuss the outcome for these patients without mention of the detail of the underlying urethral pathology [78,82,96,131,132]. Williams and Eckstein, for example, reported on 104 cases of Type I with no cases of Type III lesions [128], whereas Hendren managed to find the following combinations from 182 cases; 129 - I; one - II; 40 - III; nine - I + III; two - II + III; one - I + II [124].

There are a number points of confusion about the classification: it would appear that many have regarded the hole in the posterior urethral membrane to be a central hole, rather than at the posterior margin (e.g. Stephens in 1974); the connection to the verumontanum is often not commented on for the posterior urethral lesions; Type III and Type I lesions in the same patient usually refers to only posterior urethral lesions, rather than a bulbar Cobb's collar and posterior urethral COPUM.

The difference in the dissection technique used by Presman and Robertson was removal of
the anterior wall of the posterior urethra. Presman presented seven cases who died in infancy or early childhood, and were usually managed by suprapubic urine drainage and, thus, no urethral instrumentation; five had a posterior urethral membrane [3]. The Robertson and Hayes work involved the study of 5,622 West Indian autopsies over 15 years, in which they found 20 boys with urethral obstruction, 17 of whom had a posterior urethral membrane with a pin hole meatus on the verumontanum [5]. Others making similar observations have included Kendall and Karafin, who stated in the Journal of Urology in 1975, "it is indeed possible that a type 3 [sic] diaphragm is in reality a more extensive type 1 [sic], with the cusps continuing to meet at 12 o'clock on the roof of the urethra" [79]: Wigglesworth, in 1984 [133], in his book of neonatal pathology, shows specimens of a membranous obstruction as illustrated by Robertson and Presman, suggesting that the obstruction was not valvular, but due to a membrane. Up until these observations the classification of Young et al. had not been seriously challenged, although Kaplan stated, in 1976; "This classic system of categorisation has never been seriously challenged, yet it is not particularly useful and it may even be inaccurate" [11].

Parkkulainen [4] photographed seven boys in whom he undertook the primary treatment of urethral obstruction. He described all seven as having the characteristics of Type I and Type III in each of the patients, referring to the change from the posterior urethral membrane with a posterior hole being changed to the valve leaflets, as observed in this study, but he did not document the change with simply the passage of a urethral catheter, the Whitaker hook or the cystoscope as I have in the patients in this study (see Fig.9, 11, 24). It is interesting to note that the Kelalis and King text book picture of Type I and Type III shows the Type I lesion to have a ragged edge, similar to Fig.11, suggesting the damage to the membrane had produced the Type I appearance in their patient [10,11].

An alternative perspective was presented by Gibbons et al. [19], who argued that the bulbar urethral anomalies are congenital strictures, not the Type III posterior urethral lesions of Young, as suggested by Devereaux and Williams [134] and Stephens [18]. I would argue
that the membrane with the central hole, with no connection to the verumtanum, is bulbar pathology, of urogenital membrane origin and that this version of Type III is the same as a fibrous Cobb's collar and a congenital stricture.

My thesis reports the undisturbed anatomy of the posterior urethra as observed while assessing the efficacy of the Whitaker diathermy hook [77] and during endoscopy for fulguration. This prospective study has involved the use of preoperative suprapubic catheters and subsequent endoscopic video recording of entry into the urethra. All boys managed in this way were found to have a membranous obstruction with a pin hole orifice adjacent to the verumtanum. Furthermore, insertion of a urethral catheter, and the use of the diathermy hook converts these membranes into Young's "valve leaflets".

The cases with an obstructive membrane all had the same essential features 1) ability to prolapse down the urethra; 2) attachment of folds to the caudal end of the verumtanum; 3) more distal attachment anteriorly; 4) a 4mm defect in the posterior aspect of the membrane; 5) change of the configuration of the posterior defect with instrumentation.

As yet there does not appear to be any clinical relevance to the identification of a common morphology for most boys with congenital obstruction of the posterior urethral, although the concept of the membrane adds to the suggestion that splitting of the membrane, in those that will do, allows for partial treatment with less invasive treatment, such as Fogarty balloon [76], Whitaker hook [77] or modified valvulotome [135] disruption; this would concur with the view of Kjellberg et al. who state the "valves" become obstructive when their anterior margins fuse [136] - time and careful observation will tell.
Embryology

Kaplan, in 1976, advanced the concept that the Type I (COPUM) and Type III (Cobb's collar) should be considered separately when contemplating the embryology, with which I would agree. We also share the view that the unifying term of "posterior urethral valves" leads to confusion and misinterpretation; therefore, the discussion on embryology will be separated into the two pathologies for simplicity. To further stream-line the debate, the origin of the Type II lesion will be considered to be due to a more distal obstruction, not due to a primary embryological process.

There have been three main theories for the formation of the COPUM (Type I with anterior fusion): the structures involved in the theories related to the development of this lesion include the mesonephric (Wolffian) duct, Müller's tubercle, the sinovaginal bulbs and the urethrovaginal folds; these structures lead to the formation of the male ductal system, the verumontanum, the utricle and the plicae colliculi respectively [11,127]. Most agree on the embryology for the formation of Cobb's collar (Type III valves below the verumontanum), with the involved embryological structure being the urogenital membrane. Type III (above the verumontanum), has never been entertained as an entity since Young's final suggestion in 1929.

The embryological theories are:

1. **Exaggeration of normal folds and ridges.**

Tolmatschew attempted to explain the occurrence of the posterior urethral obstruction as due to exaggeration of normal folds and ridges [40]. Gonzales [127] and Kaplan [11] believed these folds became prominent because of a failure of posterolateral migration of the urethrovaginal folds, with anterior fusion, consistent with the lack of plicae colliculi in these patients and the tendency for the lumen to be located posteriorly. Bradford Young
quotes others as likening these inframontane folds to an "imperforate hymen" [20]. Wilckens [52], and Knox and Sprunt [56] agreed with Tolmatschew; Williams and Eckstein also agreed with the prominence of normal folds theory, but suggested that the folds may be from Wolffian structures, as suggested below [128].

2. Persistence of structures related to the incorporation of the Wolffian and Müllerian ducts into the verumontanum.

Lowsley in 1914 [41] considered that the valve-like malformations were related to defects in the development of the Wolffian and Müllerian ducts. His explanation was based on histological study of the normal and abnormal urethra in which he found the ducts in question to enter the urethra at the verumontanum, ensheathed in connective tissue, which disappeared in the urethral floor below the entry of the ducts. In Lowsley's specimen this connective tissue persisted beyond the verumontanum, gaining attachment to the walls of the urethra in such a fashion as to obstruct the lumen. His view was reiterated in 1934 [72], an opinion which was shared by Stephens [18].

3. Fusion of the colliculus with the posterior urethral groove.

In 1918 [137] and 1922 [138], Watson, working in Hugh Hampton Young's hospital, published two studies of the anatomy and development of the male urethra. The first was titled "The structure of the verumontanum - a study of the origin and development of its inherent glandular elements" and the latter, "The structural basis for congenital valve formation in the posterior urethra". Interestingly, both the papers contain the same illustration of a cross-section of the prostatic urethra of a 14 week old male fetus. Not surprisingly, Young, in both his 1919 and 1929 papers [1,2], agrees with Watson's conclusion that the anterior attachment of the verumontanum is the underlying mechanism for the formation of most posterior urethral obstructive lesions. Many subsequent authors have referred to Watson's work, without concurring with the conclusion.
4. Persistence of the urogenital membrane.

Bazy (1903), reporting on a series of six cases, suggested that the anomaly was due to persistence of the urogenital diaphragm [49]. This theory has been supported by, among others, Lederer [55], Young et al. [1], Kretschmer and Pierson [139], Kaplan [11] and Gonzales [127]. Thompson stated that "presumably this septum must be the structure formed by the cloacal hypoblast and the invaginating perineal epiblast, analogous to the anal canal and hymeneal membranes" [53]. Stephens agreed with the concept of persistence of the urogenital membrane as a cause of Type III lesions. He produced a series of diagrams indicating the canalization of the region of the membranous and bulbar urethra, with the ingrowth of the ectoderm and the down-growth of the endoderm, to explain differing patterns of the obstructive anomaly according to the manner in which the two portions meet [11]. However, his explanation does not seem to quite fit with the concept of the splitting of the cloaca to form the urogenital sinus anteriorly. Nevertheless, the persistence of the urogenital membrane would fit with the fibrous form of the inframontane lesion which I prefer to refer to as Cobb's collar, because it is bulbar in position, not valvular and not within the posterior urethra.

Based on observations of the undisturbed anatomy, the embryology of the posterior urethral obstruction would appear to be similar for most cases of congenital obstruction; a single embryological phenomenon would therefore explain the findings. The definitive evidence of bands connecting the verumontanum with the anterior urethral wall of a 14 week fetus (provided by Watson [137,138]), seems difficult to refute. Further studies of similar specimens are needed to clarify the embryology, obviously these investigations will be assisted by the new insights gained from publications from this thesis.
Variable Expression

That congenital posterior urethral obstruction is due to a membranous rather than a valvular mechanism would seem a reasonable conclusion from the above data and the literature review. The embryological mechanism for the formation of the abnormality is as yet uncertain, but the prospect of there being a variable expression of the abnormality would appear likely, given the usual divergence of manifestation for other congenital, genitourinary tract anomalies. Surprisingly, little has been published to support this view, despite the ease with which I have documented a range of abnormalities in the illustrations provided (the rarity of the minor anomalies could easily be related to the lack of clinical problems in those boys and, therefore, few needing investigation).

Detailed presentations on a broad spectrum of prominent folds running from the verumontanum have been provided by Stephens in 1955 [17], Williams in 1968 [30], Hendren in 1971 [124], Arnold and Ginsburg in 1974 [125], Kendall and Karafin in 1975 [79], and Glassberg in 1985 [140]. The most definitive study has been from Hendren who reported 182 cases of congenital urethral obstruction in patients who presented from birth to 18 years of age. He challenged previous concepts by noting that seventy-one cases did not have secondary upper tract changes, concluding that the condition is more common than previously appreciated and has a spectrum, from mild to severe. However, Hendren's conclusions differ from mine in that he suggested the obstruction is above the proximal end of the external sphincter, which is not supported by this study. He did agree that the "valve leaflet" and "membrane with a posterior hole" appearance can occur together, but he puts the "membrane with the central hole" above the sphincter, therefore in the posterior urethra, not the bulbar where it clearly is.

Arnold and Ginsburg provided evidence of folds in 25 enuretic boys, but their view and that of Hendren, is at a variance with the Paediatric Urology community because they
argue that these minor lesions cause abnormal function of the bladder (Duckett, personal communication). I would argue that wetting and urinary tract infection in these boys does not necessarily have a cause-and-effect relationship with the finding of a minimal membrane on endoscopy.

A further variation in the expression of the anomaly is the degree of the dilatation of the posterior urethra on the cystogram in the obstructive cases. In the patients investigated in this study, the degree of dilatation ranged from huge distension in Fig. 15A (similar to the cystogram presented by Cass and Stephens [93]) to minimal dilatation as seen in Fig.20 and Fig.23: a finding supported by DI Williams [30], Hendren [124] and cystogram pictures provided by Bradford Young [20] and Griesbach et al. [73]; the latter also has a cystogram of non-obstructive folds. This finding contrasts with the view of Ditchfield et al. who suggested that a lack of urethral dilatation excluded the diagnosis of obstruction, which from the above observations is untrue [141].

The findings of this study may be unique, not because of an unusual group of patients, but because of the database supported, video recorded endoscopy system.

Position of the External Sphincter

Work by Hendren in 1971 demonstrated a spectrum of the obstructing pathology, supported Young’s classification, and studied the relationship between the obstruction and the urethral sphincter, leading Hendren to suggest that the proximal part of the external sphincter is distal to the obstruction [124]. Others with a similar view include Crooks, who makes a passing comment on the obstruction being above the sphincter [130], and Popek et al., who provide an illustration with an obstruction proximal to the external sphincter. Alternatively, Stephens in one cystogram picture suggested the proximal end of the external sphincter is above the obstruction; Bradford Young, in his Fig.38, provides a line drawing
of the external sphincter overlying the folds coming from the verumtanum onto the obstruction [20] and Griesbach et al. [73], has a cystogram which probably shows the sphincter proximal to the obstruction, although they did not record it as such.

Overall, there have been few publications specifically documenting the relationship between the posterior urethral obstruction and the external sphincter and no previous study has correlated the endoscopic and radiological findings of a large group concurrently. This part of the study was to correlate the endoscopic and radiological findings in 42 cases, focusing on the nature of both obstructing and non-obstructing urethral lesions and the relationship of the membrane to the verumontanum and the external sphincter. The interpretation of the data takes into account findings from anatomical dissections of the external sphincter complex that conclude it to be a tube of muscle extending from the distal prostate to the bulbar urethra [6].

Firstly, the detailed study by Hendren should be further discussed. In 1971, Hendren reported 182 cases of congenital urethral obstruction in patients who presented from birth to 18 years of age [124]. Interestingly, Hendren reported that three of his cases had Type II lesions (above the verumtanum), and he described Type I and III lesions to be below the verumtanum but proximal to the external sphincter. My endoscopic and radiologic data clearly demonstrate that the proximal extent of the sphincter is above the level of the COPUM, whether the membrane is obstructive or merely a vestigial remnant. The cystourethrogram of Fig.6, from Hendren's 1977 publication would seem to show an indentation caused by the proximal end of the urethral sphincter and Cobb's collar, rather than the COPUM and the more distal urethral sphincter [142] and therefore the MCU concurs with my view.

Stephens studied 112 male cystourethrograms and three autopsy specimens, concluding that the proximal end of the external sphincter is located just below the verumtanum and proximal to the obstruction (not in the text, but in one of the images provided). He also
regarded the sphincter as a tube of muscle from the prostate to the urogenital diaphragm, but suggested that the bulbar muscular narrowing is due to contraction of the bulbo-cavernosus muscle rather than the distal end of the sphincter complex [17].

Part of the confusion in defining the relationship between the posterior urethral membrane and the external sphincter relates to the difference between the endoscopic and the anatomical dissection findings. Urologists identify the external sphincter as a flat plate of muscle, below the verumontanum, whereas Olerich has demonstrated the external sphincter to be a tube of muscle which, in the fetus, is between the base of the bladder and the cloacal membrane [6]. During growth of the prostate, the cranial muscle fibres are obliterated. The endoscopic appearance of the "external sphincter" as a ring of muscle of minimal length may be due to the proximal end being the physiological portion of the external sphincter complex, similar to the lower oesophageal sphincter being the active part of the oesophageal muscle. The cystogram view of the sphincter would also represent the proximal extent of the sphincter. A muscular Cobb's collar may well represent the distal extent of the sphincter tube.

Conclusion

Congenital membranes of the posterior urethra appear to have a spectrum of degree of obstruction, but for all cases, the physiological component of the external sphincter appears to be between the verumontanum and the membrane, lying on the folds that run between the two.
COPUM IMPACT ON RENAL ALLOGRAFT SURVIVAL

Introduction

It is well recognised that vesical dysfunction may continue after successful ablation of a COPUM [83,143-145]. Factors influencing bladder performance include detrusor compliance and instability, VUR and dyssynergia. In certain cases a combination of these factors will lead to poor storage capacity, incomplete emptying, and intravesical storage pressures high enough to cause progression of upper tract changes, which, if unrecognised after renal transplantation, may lead to impairment of function and graft loss. Recognition of three such cases prompted a review of the Australia and New Zealand Dialysis and Transplant Registry (ANZDATA) [146]; thus, included in this report is the largest published series of COPUM patients to receive a renal transplant and along with three case histories of graft dysfunction due to an abnormal bladder.
Materials and Methods

The ANZDATA Registry follows the clinical course and management of all patients in renal failure in 52 centres across Australasia [146]. Review of the Registry revealed 559 patients aged 20 years, or less, at the time of first renal allograft transplant between January 1975 and December 1990. Among these were 25 males whose end stage renal disease was as a result of a COPUM. The recipient age, immunosuppression, transfusion rate and donor source have been compared to the remaining 534 patients for variables recognised to influence graft survival, confirming that the two groups are similar, allowing the actuarial survival and graft survival curves to be meaningfully compared. The urethral obstruction patients were transplanted, without any attempt to standardise the surgical technique in the different units.

A detailed study of three boys who appeared to lose function of a renal graft due to bladder dysfunction was also undertaken, including a review of the biochemistry, radiology and bladder dynamic studies.
Results

The spectrum of diseases causing renal failure in the series is listed in Table 4.

<table>
<thead>
<tr>
<th>Disease</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Posterior Urethral Obstruction</td>
<td>25</td>
</tr>
<tr>
<td>Glomerulonephritis Variants</td>
<td>170</td>
</tr>
<tr>
<td>Vesicoureteric Reflux</td>
<td>149</td>
</tr>
<tr>
<td>Medullary Cystic Disease</td>
<td>44</td>
</tr>
<tr>
<td>Hypoplasia/Dysplasia</td>
<td>41</td>
</tr>
<tr>
<td>Other Obstructive Uropathy</td>
<td>22</td>
</tr>
<tr>
<td>Haemolytic Uraemic Syndrome</td>
<td>21</td>
</tr>
<tr>
<td>Alport's Syndrome</td>
<td>21</td>
</tr>
<tr>
<td>Cystinosis</td>
<td>13</td>
</tr>
<tr>
<td>Interstitial Nephritis</td>
<td>8</td>
</tr>
<tr>
<td>Polycystic Disease</td>
<td>7</td>
</tr>
<tr>
<td>Other</td>
<td>38</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>559</strong></td>
</tr>
</tbody>
</table>

Table 4: Primary renal disease in 559 renal allograft patients.

The mean age of transplantation for the COPUM patients was 14.3 years (range 4-20), compared to 15.1 years (range 1.1-20) for the remainder. Immunosuppression management is given in Table 5.

<table>
<thead>
<tr>
<th></th>
<th>Aza/Pred</th>
<th>CyA/Aza/Pred</th>
<th>CyA Only</th>
<th>Other/Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>COPUM (25)</strong></td>
<td>9 (36)</td>
<td>5 (20)</td>
<td>5 (20)</td>
<td>6 (24)</td>
</tr>
<tr>
<td><strong>Other (534)</strong></td>
<td>167 (31)</td>
<td>114 (22)</td>
<td>55 (10)</td>
<td>198 (37)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>176 (31)</td>
<td>119 (21)</td>
<td>60 (11)</td>
<td>204 (37)</td>
</tr>
</tbody>
</table>

Table 5: Post transplant immunosuppression and transplant management. The numbers in brackets () are percentages.
Transfusions preoperatively were used in 15 (60%) of the COPUM group and 385 (72%) of the remainder of the group; living-related donor transplantation was used in seven (28%) COPUM patients and 140 (26%) of the remainder of the group. There is no significant difference for any of these parameters, nor those in Table 5, using the Chi² test.

Renal allograft survival is shown in Figure 54. While the curves show some separation beyond six years post-transplant, the small number of grafts in COPUM patients remaining precludes the difference achieving statistical significance. However, three cases in which there is strong evidence for implication of bladder dysfunction in the failure of the allograft are outlined below.

**Figure 54:** Renal allograft survival for recipients less then 20 years of age, from 1975 to 1990, comparing congenital posterior urethral obstruction with the remaining population.
Case 1:

AS presented at 15 days of age with sepsis and dehydration. Investigations identified urethral obstruction which was ablated one week later. A nuclear medicine scan revealed three normal kidneys, but an intravenous pyelogram (IVP) one week later demonstrated right sided hydronephrosis. The patient's serum Cr failed to settle, and two months of age was 205 μmol/l. The patient was next admitted at five months of age with a Klebsiella urinary tract infection and a serum Cr of 500 μmol/l. His urethra was normal endoscopically and on the cystourethrogram. The infection was controlled, but gradual progression to chronic renal failure occurred, resulting in bilateral upper tract diversion at 18 months of age. However, no improvement in renal function was seen at this late stage.

The patient received a cadaveric renal transplant at two years of age from a four year old, head-injured donor. Immunosuppression with prednisolone and cyclosporine produced excellent early graft function, clinically, and a nuclear medicine scan and graft US were normal. Following the removal of an indwelling urinary catheter there was prompt deterioration of function which showed no response to anti-rejection treatment, and he developed transplant hydroureteronephrosis, despite no evidence of vesicoureteric junction obstruction. His renal function declined and the upper tract dilatation progressed until an indwelling catheter was re-inserted, with a fall in the Cr from 245 μmol/l to 174 μmol/l in a 24 hour period. A subsequent urodynamic study revealed a hyperreflexic, dyssynergic bladder with a peak pressure of 60 cm H₂O at the small total capacity of 110 ml and a residual volume of 60 ml. The patient progressed to chronic renal failure and dialysis as the significance of high bladder pressures was not recognised. He subsequently had a bladder augmentation and commenced clean intermittent catheterisation and now has a good volume, low pressure bladder, into which a second graft has been successfully implanted.
Case 2:

JF presented at two weeks of age with vomiting, poor feeding and failure to thrive. On investigation, his serum Cr was 220 μmol/l and imaging revealed bilateral hydroureteronephrosis and an obstructed urethra, a trabeculated bladder and a large urachal remnant. His differential GFR was 7 mls/min/1.73m² on the left and 11 mls/min/1.73m² on the right. His COPUM was successfully fulgurated, and his serum Cr slowly settled, then began to rise, reaching 200 μmol/l in the second month. At the age of three months, ureteric reimplants were performed for unconfirmed vesicoureteric junction obstruction, without improvement in his renal function. Dialysis was commenced at four years of age and a transplant, from his mother, was performed one month later.

His serum Cr fell to less than 30 μmol/l in the first month post transplant, then gradually increased to 100 μmol/l over the ensuing ten months, at which time a sharp rise to 182 μmol/l occurred. Further investigation with an IVP and MCU showed transplant hydronephrosis, with VUR, but no urethral obstruction. An anti-reflux reimplantation of the transplant ureter was performed with a preoperative diagnosis of obstruction, however, at operation the ureter admitted a 10FG catheter with ease, suggesting a bladder cause. There was no evidence of rejection or infection at that time. The serum Cr again settled to 80 μmol/l over the next week, coincident with post-operative catheter drainage of the bladder.

Reflux into the graft redeveloped and, on cystography, the degree of bladder trabeculation was seen to have increased. The serum Cr progressively rose to 750 μmol/l, at which time dialysis was re-commenced. After five months without immunosuppression, his transplant kidney was removed and showed changes of chronic rejection.

Following removal of his transplant kidney, he was polyuric and became incontinent. A cystogram showed an irregular bladder of moderate volume, consistent with a urodynamic
study which confirmed a non-compliant and unstable bladder with a pressure of 60 cm H₂O at a volume of 200 ml. A cystoscopy and examination under anaesthetic demonstrated his urethra to be difficult to intubate. He therefore underwent, in addition to a clam ileocystoplasty, a left nephrectomy and formation of a continent, ureteric stoma in the left iliac fossa. His post-operative urodynamic study showed a much improved bladder volume and compliance. He is continent on intermittent catheterisation, and awaiting a further transplant.

Case 3:

SC presented at 12 days of age with urinary tract infection, renal failure and metabolic acidosis. A cystogram revealed a trabeculated bladder, multiple diverticula and gross left VUR. The prostatic urethra was dilated and a urethral diverticulum was demonstrated distal to the membranous urethra, causing obstruction. An isotope renal scan showed bilateral poorly functioning, scarred kidneys. A vescostomy was established, and closed two months after urethral surgery. His renal function showed progressive deterioration, so that at the age of nine years, he underwent a living-related donor renal transplant. Pre-transplant urodynamic studies showed that he had a low pressure, good volume bladder. Subsequently, he was able to void spontaneously, with satisfactory bladder emptying, and had only intermittent wetting, which improved on anticholinergic medication. A video urodynamic study performed two years later revealed his bladder had become a high pressure unstable bladder, with native ureter VUR which was then bilateral. His urine storage capacity was 184 mls, a significant proportion of which was in the left kidney and ureter, and the degree of trabeculation had increased. A ureterocystoplasty and Teflon injection of the refluxing right ureteric orifice was therefore carried out; his bladder was thick-walled and trabeculated. On the sixth post-operative day, his transplant ureteric catheter was removed, which was followed by an increase in his Cr from 80 to 170 μmol/l over the next 12 hours, associated with the development of hydronephrosis. No other
cause for graft dysfunction was found, and the Cr settled by re-commencing his anticholinergic medication. He then had a good urine storage organ, and was dry without anticholinergics, but with inefficient bladder emptying. His Cr gradually increased to 140 μmol/l over the next 10 months, accompanied by intermittent moderate hydronephrosis and significant residual bladder volumes. It was felt he needed to improve his bladder emptying and, because of the combination of failed conservative management and the presence of a urethral diverticulum preventing intermittent catheterisation, an intubatable, ureteric stoma was formed from his native, right ureter. Post-operatively, his Cr came down to 80 μmol/l where it has stayed.
Discussion

An important distinction between adult and paediatric patients who enter renal dialysis and transplant programs is the much greater proportion of children who have a urological cause for renal failure [147]. In some instances urological abnormalities may persist and pose a threat to the transplanted kidney [148]. Patients with a COPUM, for example, may display evidence of continuing bladder dysfunction many years after successful ablation of the obstruction [83,143-145] and are at risk of developing secondary impairment of renal function. Case 1 and Case 2 were treated successfully for their COPUM but continued to progress to chronic renal failure and transplantation, only to lose their grafts, with evidence pointing to bladder dysfunction as the cause of the graft loss. Case 3 had graft loss prevented because of the recognition of progressive bladder dysfunction. These three cases provoked a review of ANZDATA in an attempt to clarify the problem further.

The 25 patients on the ANZDATA Registry who entered chronic renal failure as the result of COPUM, and were transplanted, are similar to all other patients transplanted over the same period with respect to recipient age, immunosuppressive regimen, transfusion treatment and donor source. The comparison of allograft survival of the two groups should not, therefore, be influenced by these variables. The actuarial graft survival for COPUM patients was not significantly different to the remainder of the similarly aged patients, as found by others [149]. Although the analysis of the ANZDATA suggests that transplantation into a post-COPUM bladder is a safe option, these three cases demonstrate that this is not always the case, and the similar graft outcome for both groups from the ANZDATA may be related to the incidence of severely dysfunctional bladders within the group of COPUM patients being small, thus a large series would be required to achieve a statistically significant difference in renal transplant outcome. Interestingly, the three cases were younger than mean age at transplantation of the ANZDATA series, and perhaps renal failure at a young age may be related to more severe bladder dysfunction. Therefore, patients treated for a COPUM who progress to chronic renal failure and transplantation at a
young age should be followed closely, and assessment of adverse changes in the transplant kidney should include consideration of bladder deterioration as a possible cause. The most appropriate test is a urodynamic study, usually through a suprapubic catheter, during which the volume, end-fill pressure, bladder instability and compliance should be assessed. If the bladder is sufficiently implicated, then, depending on the nature of the bladder findings, anticholinergic medication or bladder augmentation may be required.
COBB'S COLLAR

Introduction

I would suggest a classification of Cobb's collar which incorporates the degree of encroachment on the lumen and the presence of muscle or a fibrous membrane within the narrowed segment. This suggestion is founded on a computer database assisted review of video tapes of 210 boys endoscoped on 346 occasions over a five year period, 119 of whom had a suitably detailed recording of their bulbar urethra. The minor Cobb's collars were not always seen on cystogram, and are therefore not of pathological importance, but the study of such lesions may help improve the understanding of the development of this part of the male urethra.

Stephens [17,18], Gibbons et al. [19] and Currarino [14] suggested that indentation on the cystogram can be due to a separate muscle structure, however the narrowing in most cases seems better related to the lower end of a tubular sphincter, particularly as the constriction endoscopically appears circumferential. The fibrous narrowing at the bulbar urethra has been variously termed Cobb's collar, Moormann's ring, congenital stricture and the original concept of Type IIIb by Young et al. [1]; where there is a bulbar urethral membrane with no connection to the verumontanum, the pathology and the embryology is probably all the same.
History of Cobb's collar

Cobb et al., in 1968, [16] were the first to describe a significant number of cases of narrowing in the bulbar urethra which appeared to be congenital; hence the term Cobb's collar. They suggested that minor changes could often be seen if the urethra was entered under direct vision. However, the understanding of Cobb's collar appears to have been confused by studies grouping boys with older men [150-153], studies of urethrograms without cystoscopy [14], confusion between prolapsed lesions of the posterior urethra and those which are of the bulbar urethra primarily [75,154] and regarding all bulbar constrictions as pathological rather than some as merely anatomical variations [155]. The term Moormann's ring [152] is also used, because of the study published in 1972, for the same bulbar urethral pathology, and the presence of two separate terms may have added to the confusion. Others have also suggested that primary congenital obstructive pathology can occur in the bulbar urethra, [3,156,157] but none have been as convincing as the presentation by Cobb et al. [16].
Materials and Methods

Over a five year period from October 1990 to October 1995, 210 boys had a video recording, of which 66 were found to have a localized constriction in the bulbar urethra. The videos were reviewed with the assistance of a computer database. Where an adequate view of the urethra was not recorded during a single endoscopy, an alternative video was used in those boys who had more than one procedure, giving 178 boys in whom a satisfactory view of the bulbar urethra was obtained. Of the thirty-two cases excluded from the study, in 15 the anatomy was not adequately visualized, 16 cases had treatment or grossly abnormal anatomy, and one was excluded because the lesion was thought to be a prolapsed more proximal membrane because of attachments to the verumontanum.

In the 66 cases (aged 55.7±49.3 months - Fig.55) with a narrowing below the external sphincter, the nature of the lesion was noted, in particularly any variation in the calibre of the narrowing during the examination, its relationship to the external sphincter and the presence of other urethral pathology. The cystogram of the 54 boys who had satisfactory urethral views were also studied, and evidence of a bulbar urethral narrowing separate to the external sphincter documented.

Figure 55: The graph of the age distribution for the 66 boys with a Cobb's collar on endoscopy.
Cobb's collar

Results

Of the 66, five had a significant narrowing, 34 had a moderate change in the lumenal diameter, and 27 had a minor indentation. Eight boys had a membranous obstruction, 46 had a lesion which changed appearance during the endoscopy, consistent with contracting muscle, and 12 (all minor abnormalities) had a constriction of uncertain nature. Seventeen boys also had an additional more proximal membrane in the posterior urethra.

In the 54 boys with a Cobb's collar, who had a satisfactory view of their bulbar urethra recorded during the preoperative urethrogram, 41 were found to have an indentation in the bulbar urethra, separate from the external sphincter, which correlated in position with the cystoscopic findings. Most of the bulbar urethral lesions were an incidental finding.

The results have been grouped according to the degree of impingement on the urethral lumen and the nature of the pathology as suggested by any alteration in the appearance during the endoscopy in Table 6, Table 7 shows the cystogram findings and Table 8 the presentation of the 66 boys.


<table>
<thead>
<tr>
<th></th>
<th>Muscular</th>
<th>Membranous</th>
<th>Unknown</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Minimal</td>
<td>15</td>
<td>-</td>
<td>12</td>
<td>27</td>
</tr>
<tr>
<td>Moderate</td>
<td>29</td>
<td>5</td>
<td>-</td>
<td>34</td>
</tr>
<tr>
<td>Severe</td>
<td>2</td>
<td>3</td>
<td>-</td>
<td>5</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>46</td>
<td>8</td>
<td>12</td>
<td>66</td>
</tr>
</tbody>
</table>

Table 6: Bulbar urethral abnormalities grouped in type and degree, according to the endoscopic findings.

<table>
<thead>
<tr>
<th></th>
<th>No MCU</th>
<th>Not seen</th>
<th>Min.</th>
<th>Mod.</th>
<th>Def.</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Minimal</td>
<td>8</td>
<td>9</td>
<td>5</td>
<td>1</td>
<td>4</td>
<td>27</td>
</tr>
<tr>
<td>Moderate</td>
<td>4</td>
<td>4</td>
<td>10</td>
<td>5</td>
<td>11</td>
<td>34</td>
</tr>
<tr>
<td>Severe</td>
<td>-</td>
<td>-</td>
<td>1</td>
<td>4</td>
<td></td>
<td>5</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>12</td>
<td>13</td>
<td>15</td>
<td>7</td>
<td>19</td>
<td>66</td>
</tr>
</tbody>
</table>

Table 7: Cystogram findings of the 66 boys with a Cobb's collar, matched to the degree of observed narrowing on endoscopy. Min. - minimal; Mod. - moderate; Def. - Definite.
Table 8: Clinical presentation groups.

<table>
<thead>
<tr>
<th>Condition</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Urinary tract infection</td>
<td>29</td>
</tr>
<tr>
<td>Haematuria</td>
<td>7</td>
</tr>
<tr>
<td>Prenatal diagnosis</td>
<td>13</td>
</tr>
<tr>
<td>Incontinence</td>
<td>8</td>
</tr>
<tr>
<td>Poor stream/retention</td>
<td>3</td>
</tr>
<tr>
<td>Penile pain</td>
<td>2</td>
</tr>
<tr>
<td>Other</td>
<td>4</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>66</strong></td>
</tr>
</tbody>
</table>

Figure 56 shows a minor indentation, Figure 57 a moderate degree of lumenal change with the associated cystogram, Figure 58 is the cystogram and endoscopic view of a membranous obstruction; Figure 59 shows a severe obstruction caused by the membranous lesion seen in Figure 62; an obstructed retrograde study is seen in Figure 61, which is seen endoscopically in Figure 60. Figure 63 shows a more proximal view through the obstruction, indicating the external sphincter is separate from the bulbar lesion.

Figure 56: A minor indentation seen in the bulbar urethra, separate from the external sphincter seen more proximally.
Figure 57A: A cystogram showing the indentation from a moderate muscular Cobb’s collar seen endoscopically in Fig. 57B.

Figure 57B: A similar view to that seen in Fig.56, from a boy with a greater degree of encroachment of the lumen. The more proximal sphincter is not shown.
Figure 58A: A cystogram of a moderate fibrous Cobb's collar seen endoscopically.

Figure 58B: Another boy with moderate fibrous encroachment of the bulbar lumen. The proximal extent of the sphincter not is shown, but the associated syringocele is seen.
Figure 59: The cystogram of the endoscopic view shown below. There is a tight stenosis in the bulbar urethra.

Figure 60: A severe membranous narrowing in the bulbar urethra, consisting of a fibrous membrane with a central hole.
Figure 61: The obstructed retrograde attempt at a cystogram of the endoscopic view shown below.

Figure 62: A severe muscular narrowing in the bulbar urethra; note the crenation, consistent with muscle.
Variation in the lumenal diameter and crenation of the overlying mucosa were assumed to imply the presence of muscle and occurred in 46 boys. In all cases the bulbar narrowing was seen to be separate from the proximal end of the external sphincter.

Seventeen boys had both a COPUM and a bulbar urethral narrowing; six of the COPUM's were minor membranes, six moderate and five obstructive.
Cobb's collar

Discussion

Congenital obstruction of the bulbar urethra has not often been discussed in the literature, possibly because many would agree with Cobb's observation in 1968 [16], that insertion of the cystoscope, whilst visualizing the urethra, will often show the presence of a ring narrowing of the bulbar urethra, and therefore it is not worthy of recording; a conclusion also reached by Cranston et al. [155]. In my patients, a Cobb's collar was found in only 66 of the 178 (37%) in whom there was an adequate recording of the bulbar urethra. However, it would seem that narrowing in the bulbar urethra is often congenital in nature and possibly due to an embryological remnant of the urogenital membrane, which is either muscular or fibrous, minor or significant. And as significant pathology does occur, the abnormality should be further clarified.

Patients with a congenital narrowing in the bulbar urethra have often been grouped with boys who have traumatic or iatrogenic strictures, leading Harshman et al. [158] to conclude that the term congenital stricture should be avoided. Never-the-less, congenital narrowing has been seen in this study and in the six cases reported by Currarino and Stephens [159], where cystograms similar to my cases were shown. The presence of a membranous lesion, where the diagnosis of hydronephrosis was made prenatally, and the young age of most of the boys, supports the notion that these lesions are congenital. This is also suggested in other studies where young boys have presented with upper tract changes with an obstructing membrane in the bulbar urethra [14,159].

Moormann's ring is another title used for a congenital narrowing in the bulbar urethra, but one wonders about the congenital nature of the patients reported by Moormann, as they were all between 21 and 54 years of age [152]. He did, however, provide endoscopic pictures similar to those shown in Figure 56 + 57.
Others have reported the familial incidence of a bulbar urethra narrowing in a father and son [150] and in brothers [151,160]. These constricting bulbar urethral lesions, in older men, may be due to spasm of intrinsic urethral muscle or spasm of the bulbospongiosus, highlighting that the bulbar urethral findings may be either muscular or membranous, and the delayed presentation of adults suggests that the narrowing is most likely to be muscular in the older patients. Currarino [14] showed a Cobb's collar impression on radiographs, with variable appearance between patients concluding that the radiological findings were due to contraction of periurethral muscle. This is in keeping with the finding of a decrease in the diameter of the constriction during the endoscopy (as if due to muscle contraction) in 46 boys. In contrast, Cobb thought the lesion could not be muscle, as the 26 children he recorded had a narrowing which was not affected by succinylcholine [16]. It can be argued that not all are muscular, but many appear to be.

The bulbar urethral lesion, due to local pathology, can be differentiated from a prolapsed COPUM by the absence of an attachment to the verumontanum, which is evidenced by 17 boys in this study, and five boys in Cobb's et al.'s paper [16], where both lesions were seen concurrently (Fig. 59).

The incidence of Cobb's collar may be less in the general community than in this study, as these 66 boys are obviously a biased population and the incidence of significant pathology is even less, as only 76% of those with endoscopic evidence of a narrowing had any abnormality detected on the cystogram. Even-so, the recognition of the nature of bulbar urethral anomalies is important for the small proportion of cases in which the pathology is significant.
lesions in 195 boys, and thus a similar incidence.

Fenwick (1896) was the first to use an endoscope to make the diagnosis. He recorded ten cases with a retention cyst of Cowper's gland, most of which may have been secondary to gonococcal infection. He performed repeat examinations using an aëro-urethroscope, described endoscopic deroofing and an association with "organic strictures of the membranous urethra". He assumed the strictures to be secondary to inflammation in the underlying cystic structure [165].

The term, syringocele, now widely used, was popularised by Maizels et al. [166]. They provided a four part classification and suggested the gland and duct to be associated with the circular external sphincter. They made no mention of Cobb's collar [16]. However, Van Brien et al. present a cystogram with the appearance of a Cobb's collar in one of their five cases [167], Yaffe and Zissin found a stricture of the bulbar urethra in six of eleven cases [168] and the cystogram of the patient reported by Dhillon et al. had a bulbar urethral narrowing [169]. The presence of both the external sphincter and muscular Cobb's collar may be explained by previous anatomical studies of the proximal urethra. Oelrich performed detailed dissections of the urethral sphincter at various ages, concluding that the sphincter is a continuous tube of muscle from the bladder to the perineum with, perhaps, a relative condensation of fibres caused by the growth of the prostate gland [6], giving the impression of a flat plate sphincter below the prostate endoscopically; an increased number of fibres at the distal end of this tube of muscle may account for the appearance of the muscular Cobb's collar in the bulbar urethra [16]. This suggestion is supported by the diagrams of Johnson's fetal dissections in 1920 [7].
Materials and methods

Eleven boys presented between October 1990 and October 1995, ranging in age from newborn to 17 years (mean 3.9 years - six boys were less than six months old).

The clinical presentations of the boys were as in the table below.

<table>
<thead>
<tr>
<th>Condition</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prenatal Hydronephrosis</td>
<td>4</td>
</tr>
<tr>
<td>Poor Stream</td>
<td>3</td>
</tr>
<tr>
<td>Urine Infection</td>
<td>3</td>
</tr>
<tr>
<td>Wetting</td>
<td>1</td>
</tr>
</tbody>
</table>

Table 9: Presentation of boys with a syringocele.

A cystourethrogram was performed in 10 boys, and endoscopy in nine. One boy had prenatal mild hydronephrosis, died at birth of unknown cause, and had an autopsy.

An oblique view of the urethra was obtained for all cystograms. These were reviewed at the time of the endoscopy, and later with the video tape of the endoscopy, in the nine boys who had both procedures.

During the endoscopy the first passage of the urethra was recorded, as were subsequent views, where appropriate. Video footage of the intervention and its outcome were also recorded. The videos of all patients were subsequently reviewed concurrently and the nature of each syringocele was noted, particularly the relationship to any bulbar urethral narrowing and the external sphincter. Re-
endoscopy was carried out in those in whom intervention was performed at the first procedure, during which the relationship of the landmarks was restudied.

**Results**

The neonatal autopsy case had bilateral, thin walled intralumenal cysts, without any evidence of bulbar urethral narrowing and no signs of urinary tract obstruction, although, a cystogram, via a suprapubic stab, identified grade III VUR.

**Cystogram findings**

Two boys had a minimal tract seen on cystogram (Fig. 64), four had a cystic filling defect (Fig. 65), and three had a diverticulum (Fig. 66).

![Figure 64: A cystogram showing a minimal tract of contrast into the Cowper's duct.](image)
Figure 65: A cystogram showing a cystic filling defect in the floor of the urethra and minimal, tapered bulbar urethral narrowing.

Figure 66: A cystogram showing a diverticulum and the Cobb's collar.
Four had no evidence of a bulbar urethral narrowing (Fig. 67), two had a minimal lumenal change and four had a moderate change.

Figure 67: Cystogram showing the calibre change in the bulbar urethra from a narrowing which appeared to be muscular on the subsequent endoscopy. The minor indentation of the proximal end of the external urethral sphincter is also seen.

The interrelationship of the syringocele and Cobb's collar findings is given in Table 10, below.

<table>
<thead>
<tr>
<th>Name</th>
<th>Cobb's</th>
<th>Syringocele</th>
</tr>
</thead>
<tbody>
<tr>
<td>DR</td>
<td>Nil</td>
<td>Cyst - minimal</td>
</tr>
<tr>
<td>CR</td>
<td>Nil</td>
<td>Cyst - moderate</td>
</tr>
<tr>
<td>SM</td>
<td>Nil</td>
<td>Tract - minimal</td>
</tr>
<tr>
<td>JW</td>
<td>Nil</td>
<td>Diverticulum</td>
</tr>
<tr>
<td>JC</td>
<td>Minimal</td>
<td>Cyst - moderate</td>
</tr>
<tr>
<td>SS</td>
<td>Minimal</td>
<td>Tract - minimal</td>
</tr>
<tr>
<td>SB</td>
<td>Moderate</td>
<td>Cyst - minimal</td>
</tr>
<tr>
<td>JA</td>
<td>Moderate</td>
<td>Cyst - moderate</td>
</tr>
<tr>
<td>DT</td>
<td>Moderate</td>
<td>Diverticulum</td>
</tr>
<tr>
<td>RS</td>
<td>Moderate</td>
<td>Diverticulum</td>
</tr>
</tbody>
</table>

Table 10: Cobb's collar and syringocele findings on cystogram for each of 10 boys.
Endoscopy findings

Six of the nine syringoceles opened into the floor of the bulbar urethra with a single opening (Fig. 68). The wall between the urethra and the tract was a fibrous membrane. In one case (SM), there was only a minimal tract. Two boys had closed cysts in the floor of the bulbar and proximal penile urethra, one of whom had bilateral syringoceles (Fig. 68B). One boy (CR) had the appearance suggestive of a ruptured intralumenal cyst.

The Cobb's collar and syringocele findings on endoscopy are detailed in Table 11, below.

<table>
<thead>
<tr>
<th>Name</th>
<th>Cobb's</th>
<th>Syringocele</th>
</tr>
</thead>
<tbody>
<tr>
<td>JW</td>
<td>Nil</td>
<td>Open distally</td>
</tr>
<tr>
<td>DR</td>
<td>Nil</td>
<td>Open distally</td>
</tr>
<tr>
<td>JC</td>
<td>Fibrous - moderate</td>
<td>Open distally</td>
</tr>
<tr>
<td>SM</td>
<td>Fibrous - moderate</td>
<td>Minimal Tract</td>
</tr>
<tr>
<td>DT</td>
<td>Fibrous - severe</td>
<td>Open distally</td>
</tr>
<tr>
<td>RS</td>
<td>Muscle - minimal</td>
<td>Open distally</td>
</tr>
<tr>
<td>JA</td>
<td>Muscle - moderate</td>
<td>Closed - double</td>
</tr>
<tr>
<td>SB</td>
<td>Muscle - moderate</td>
<td>Closed - single</td>
</tr>
<tr>
<td>CR</td>
<td>Muscle - moderate</td>
<td>Open cavity</td>
</tr>
</tbody>
</table>

Table 11: Cobb’s collar and syringocele findings on endoscopy for each of 9 boys.

Two boys had no narrowing in the bulbar urethra, one had minimal impingement, four had a moderate decrease in the lumen (Fig. 68B), and one had a marked decrease in diameter, with what appeared to be a significant fibrous narrowing (Fig. 68A), shown over the page.
Figure 68A: An endoscopic view of the urethral opening of the most common appearance of the syringoceles with a distal open cavity and a bulbar urethral narrowing.

Figure 68B: An endoscopic view of the urethral opening of bilateral closed syringoceles and a muscular Cobb's collar.
The syringocele appeared to end just proximal to the bulbar narrowing and the sphincter was seen to be separate from this narrowing in all cases (Fig. 69).

Figure 69: The proximal extent of the external sphincter can be seen above, and through the muscular narrowing in the bulbar urethra.

The cystogram and endoscopy appearance of the syringoceles appeared to concur in all except three cases. In these cases, the cyst seen on the radiological study was a distally opened cavity or a minimal intralumenal cyst wall remnant (Table 12), suggesting that the cystic dilatation had either partially or completely ruptured prior to the endoscopy.

<table>
<thead>
<tr>
<th>Name</th>
<th>Cystogram</th>
<th>Endoscopy</th>
</tr>
</thead>
<tbody>
<tr>
<td>CR</td>
<td>Cyst - moderate</td>
<td>Open cavity</td>
</tr>
<tr>
<td>DR</td>
<td>Cyst - minimal</td>
<td>Open distally</td>
</tr>
<tr>
<td>DT</td>
<td>Diverticulum</td>
<td>Open distally</td>
</tr>
<tr>
<td>JA</td>
<td>Cyst - moderate</td>
<td>Closed - *2</td>
</tr>
<tr>
<td>JC</td>
<td>Cyst - moderate</td>
<td>Open distally</td>
</tr>
<tr>
<td>IW</td>
<td>Diverticulum</td>
<td>Open distally</td>
</tr>
<tr>
<td>RS</td>
<td>Diverticulum</td>
<td>Open distally</td>
</tr>
<tr>
<td>SB</td>
<td>Cyst - minimal</td>
<td>Closed - *1</td>
</tr>
<tr>
<td>SM</td>
<td>Tract - minimal</td>
<td>Minimal Tract</td>
</tr>
</tbody>
</table>

Table 12: Syringocele findings on cystogram and endoscopy for each of 9 boys. Those with poor correlation are in bold italics.
Syringocele - Results

The cystogram showed less bulbar urethral narrowing than the endoscopy in four cases, and more in one (Table 13). The muscular lesions may have had a different degree of spasm for each study, and the thin fibrous narrowing may have been less evident on the cystogram because of contrast on either side of the membrane obscuring it.

<table>
<thead>
<tr>
<th>Name</th>
<th>Cystogram</th>
<th>Endoscopy</th>
</tr>
</thead>
<tbody>
<tr>
<td>CR</td>
<td>Nil</td>
<td>Muscle - moderate</td>
</tr>
<tr>
<td>DR</td>
<td>Nil</td>
<td>Nil</td>
</tr>
<tr>
<td>DT</td>
<td>Moderate</td>
<td>Fibrous - severe</td>
</tr>
<tr>
<td>JA</td>
<td>Moderate</td>
<td>Muscle - moderate</td>
</tr>
<tr>
<td>JC</td>
<td>Minimal</td>
<td>Fibrous - moderate</td>
</tr>
<tr>
<td>JW</td>
<td>Nil</td>
<td>Nil</td>
</tr>
<tr>
<td>RS</td>
<td>Moderate</td>
<td>Muscle - minimal</td>
</tr>
<tr>
<td>SB</td>
<td>Moderate</td>
<td>Muscle - moderate</td>
</tr>
<tr>
<td>SM</td>
<td>Nil</td>
<td>Fibrous - moderate</td>
</tr>
</tbody>
</table>

Table 13: Cobb's collar on cystogram and endoscopy for each of 9 boys. Those with poor correlation are printed in bold italics.
Discussion

Cowper's glands are analogous to Bartholin's glands in the female; they are paired pea sized structures, which arise in the third month of fetal life, as ectodermal buds, at the site of the eventual bulbocavernous urethra. The ducts grow backward and Cowper's glands become imbedded between the leaves of the urogenital diaphragm. There are often more distal, accessory paired glands [170,171]. A cystic dilatation of the duct and gland can be a congenital or acquired disease [7,165,172-174], usually presenting with urinary tract infection or haematuria [171,173,175,176]; alternatively the presence of a syringocele may be identified because of urethral obstruction [172,174,177] or a perineal mass [171,178,179]. Investigation of prenatal hydronephrosis and urinary infection were the common presenting features in the current group.

Syringoceles were considered to be sufficiently rare in 1929 to warrant reporting as a single case [178], whereas by 1978 they were considered to be more common and easily treatable by endoscopic fulguration [180]. Others have concurred with the ease of treatment endoscopically [175,179,181,182] even if the lesion presents as a perineal mass [179], whereas the perineal lesions had previously been managed by open resection [178]. However, not all syringoceles require treatment. Two of eleven Cowper's duct abnormalities in this group did not have any treatment (all operative cases were treated via an endoscope), whereas Bethencourt et al. treated only six of their 14 patients [181]. Currarino and Fuqua, in a review of cystograms, included pictures of eight of the 10 cystograms performed, all of which showed only a minor tract; the Cowper's duct anomaly could not be visualised in four of the six who had endoscopy [176]. A cystic lesion was not found in any of their patients. Furthermore, Moskowitz et al. mentioned the possibility of spontaneous decompression [173], suggesting such cases did not require further intervention. Three of the current patients had at least partial decompression,
however, fulguration of remnants of the syringocele was performed. The difference between the minimal tract and the cystic lesion would appear to be variable expression of the same embryological abnormality, with the difference between the cystogram and cystoscopic findings may be due to a combination of a change with time and/or the different perspective between the visualisation modes; the main variation in the syringocele appearances was that a cyst on cystogram was viewed as an open cavity in three cases, and a bilateral syringocele was not appreciated as double, on the cystogram (Table 12). The variation in findings between the two modalities seems to be neither significant nor surprising.

Maizels et al. provided a classification for syringoceles, the diagrams of which suggest that the proximal Cowper's duct was closely associated with the external sphincter (Fig.70) [166]. They did not identify any intrinsic urethral narrowing, but found that deroofing the cysts often failed to resolve the voiding dysfunction, which might imply that partially obstructing lesions in the bulbar urethra remained unrecognised at the time of their initial endoscopy. Given the association of strictures with syringoceles by others [165,168], and the presence of a fibrous or muscular Cobb's collar in seven of the nine boys endoscoped in this study, it would appear that syringoceles and a Cobb's collar often co-exist as shown in Figure 70, in keeping with the suggestion that both Cobb's collar and Cowper's gland arise from the region of the urogenital membrane [159]. Currarino and Stephens used the term congenital stricture, but, as it would seem that either a muscular or fibrous narrowing can be seen, the term Cobb's collar may be more appropriate [16].
Figure 70A: An imperforate syringocele and the external sphincter as presented by Maizels et al. [166].

Figure 70B: The relationship of Cobb's collar, syringocele and the physiological external urethral sphincter suggested from this study.
The muscular nature of the Cobb's collar in four cases would partly explain the different conclusions of this paper and other authors, and the different cystogram and cystoscopy findings in two of the boys in this study. However, four cases had similar findings on the cystogram and the endoscopy; the remaining three had a fibrous lesion which appeared less evident on the cystogram than on the endoscopic visualisation, which may be explained by the density of the contrast obscuring the thinner central part of the obstructing membrane.

Overall, the evidence of this study would appear to support the common association between syringoceles and Cobb's collar, suggesting a common origin, and the need to specifically exclude an associated obstructing lesion in a boy with a syringocele.
IDIOPATHIC URETHRITIS IN THE ADOLESCENT MALE

Introduction

Bulbar urethritis in adolescents is not an uncommon urological problem but its aetiology remains obscure and the literature on the subject is sparse, and usually not specifically on bulbar urethritis [134,158,183-185]. Kaplan and Brock coined the term urethrorrhagia to describe the blood spotting typically present between episodes of micturition [184]. They cautioned against the use of cystoscopy, suggesting that it may contribute to the development of a urethral stricture [183], which was at variance with my own view and, therefore a stimulus to collect and collate the data presented in this chapter. Inclusion of the observations on urethritis pertains to the disease as a cause of urethral narrowing in young boys, highlighting the need for caution when considering whether or not a lesion of the male urethra is congenital or not.

Urethral inflammation is known to be associated with immunological and infective conditions, such that urethritis may be the only presenting symptom of Reiter's syndrome [186], Chlamydia is a common cause in older men [187] and it is common in the hypogammaglobulinaemic state, often with Mycoplasma infection [188]. These associations with adolescent urethritis have not been well studied, but were researched in this group of patients by way of a literature review.

In order to further elucidate this condition all patients over a five year period with the symptom complex were extensively investigated. The stricture development, both prior to and after initial treatment was noted, particularly because of the confusion between congenital and acquired narrowing of the bulbar urethra.
Materials and Methods

Seven adolescent males, from 12-15 years old, presented with urethritis over the five year period.

All patients had urine microscopy and culture, as well as a urethral swab for Chlamydia isolation. An MCU, renal tract US and cystoscopy were performed in all patients. Appropriate segments of urethrosopic video were recorded, especially as the urethra was first entered. Two patients had urethral biopsies.

Full blood examination, HLA B27, and immune function studies were performed, including IgG and subclasses, ASOT, antiDNAse B, antitetanus and antimeasles antibodies and isoohaemagglutinins.

All clinical data, investigation results and cystoscopic video footage were reviewed.
Results

Five boys had blood spotting on the underwear, four had dysuria, and only two had macroscopic haematuria. They presented with a symptom duration of 1-6 months. Two boys had a history of minor scrotal trauma which was recounted retrospectively, but had not precipitated the referral. There was no history of either urethral instrumentation or sexual contact on direct questioning of either the parents or adolescents. All patients had empirical courses of antibiotics prior to urological review with no alteration in their clinical state. One patient, who presented with symptoms of urethral narrowing, had cystoscopy prior to referral.

Clinical examination was normal in all patients except for one with a biopsy proven drug-related skin rash, due to Cotrimoxazole. No patient had any evidence of uveitis, conjunctivitis or arthritis on either history or examination.

In all cases urinalysis revealed white and red cells but no organisms on culture of either urine or urethral swabs, including cultures looking for fastidious organisms.

All patients had a normal full blood count and erythrocyte sedimentation rate, and were HLA B27 negative. Low immunoglobulin G1 was noted in two out of seven boys tested. The other markers of antibody formation were normal.

On cystourethrogram, three patients had a minor degree of unilateral VUR (grade 1 or 2), one patient had an irregularity noted in the bulbar urethra and one had a bulbar urethral stricture. The pelvicalyceal systems were normal on all US examinations, with one patient being noted to have a solitary kidney.

The endoscopic findings were similar for the location of the inflammatory change and most had a clear cut transition from normal to abnormal. The area of
circumferential inflammation was found in the bulbar urethra (Fig. 71). At initial presentation two patients had bulbar urethral strictures requiring dilatation and two others subsequently developed narrowing which required dilatation (Fig. 72).

Figure 71: The inflamed bulbar urethra at the junction with the normal distal urethra. The pale, fibrotic, inflamed region is proximal to the more normal urethra which has only slightly increased vascular markings.

Figure 72: A narrow region in the bulbar urethra at the distal extent of a centimetre length of marked inflammatory change.
Urethritis in Adolescents

The two urethral biopsies showed non-specific inflammation with no organisms seen on microscopy or grown in culture.

Symptoms persisted for 10 months to three years and four boys have on-going symptoms.

Discussion

Bulbar urethritis in adolescent boys is well recognised but its pathogenesis is little understood and its relationship to stricture formation uncertain.

Kaplan and Brock coined the term urethrorrhagia in their series of 21 patients over an 11 year period [184]. This term was used to describe the blood spotting on underpants typically present between episodes of micturition. Six of their patients had associated dysuria and seven had protracted symptoms. Endoscopy was performed in 15 patients and an area of inflammation was described at the external sphincter and verumontanum with an ulcer in the bulbar urethra in one patient. Whereas, in my study, patients had an inflamed area in the bulbar urethra, separate from the external sphincter, with ulceration in all cases. The disease may be different in the two groups or the observations less reliable when videos of all cases are not viewed concurrently, as they were in this group. Three of the patients of Kaplan and Brock subsequently developed urethral strictures, leading the authors to suggest that cystoscopy may have led to the development of these strictures. They felt endoscopy in this symptom complex was unnecessary and perhaps detrimental. Williams and Mikhael presented 17 cases and described bulbar urethritis in only 11 on cystoscopy [185]; five had some hyperaemia of the posterior urethra, one was normal and two were found to have bulbar urethral strictures. This was more in keeping with the current patients in whom the incidence of bulbar urethral strictures
was high [4 of 7 patients] and two of these strictures were found at the initial cystoscopy. It would appear that stricture formation is a result of the disease, not the intervention, and therefore, the condition should be managed accordingly, which concurs with the view of Williams and Mikhail [185].

The aetiology of bulbar urethritis is not known. It is possibly an inflammatory urethritis as seen in Reiter's syndrome, but without the uveitis or arthritis typical of Reiter's. The cause of the urethritis in Reiter's is not known but presumably it represents an autoimmune process. All of my patients were carefully evaluated for other evidence of Reiter's syndrome, none of which was found. All were negative for HLA B27. In a review of 26 patients (85% were male) with juvenile onset Reiter's syndrome the mean age at onset was 10.5 years [186]. The patients presented with urethritis alone in 23%, all subsequently developed arthritis which was mainly lower limb and 69% pauciarticular. Conjunctivitis developed in 88%, urethritis in 54% and 67% were HLA B27 positive. Reiter's, therefore, should be considered a diagnostic possibility in the adolescent with blood spotting on his underwear, but was not diagnosed in the group of boys studied by myself.

Another possible aetiology is an as yet unidentified infective agent. In a cohort of 112 men presenting with non-gonococcal urethritis over an eight month period to two major centres, Chlamydia was detected in only 49% of patients despite an extensive search [187]. Chlamydia was sought by direct immunofluorescence of urethral smears, direct immunofluorescence, IDEIA and polymerase chain reaction of the centrifuged deposit of first-pass urine. Clinically there were no differences between the Chlamydia positive and negative groups. The authors conclude that in a proportion of men it may be that the organism has been cleared or that the elementary bodies have been shed to a level below that which is detectable. Further study of a possible infective agent in the adolescent age group is therefore indicated.
Immunological testing was instigated in my patients, because of a suggested relationship between hypogammaglobulinaemia, urethritis and Ureaplasma Urealyticum infection [188]. Also, Mycoplasma and Ureaplasma are known causal agents of arthritis and genitourinary infections in the hypogammaglobulinaemic population [188]. In a series of female children analysed for Ureaplasma and Mycoplasma colonisation, to determine if their presence was related to sexual contact, it was concluded that there was significant carriage of these organisms unrelated to sexual activity [189]. To my knowledge this information is not available in male children. The culture technique for Mycoplasma and Ureaplasma is now well developed, but was not attempted at the time of presentation of the seven patients. It is planned to prospectively evaluate future patients for Ureaplasma infection.

Two of seven boys tested had low IgG but as their ability to produce antibodies seemed to be maintained, the significance of this result is not clear. It would be interesting to prospectively collect data on immune status as the level of IgG may alter during the course of the disease. If consistently low levels of IgG are found there may be a place for immunoglobulin therapy which has been recommended for therapy of Ureaplasma infection associated with hypogammaglobulinaemia [190].

As the associated finding is Mycoplasma infection, treatment with Erythromycin or its newer analogue Roxithromycin would be appropriate therapy in childhood. However, Erythromycin was used in three of these boys without success, the reasons for which are either a resistant organism or the infection/inflammation is not responsive to the antibiotic.

Urethritis may also be due to gonococcal infection or auto-instrumentation. There was no history of intercourse, urethral swabs were negative, the boys all denied instrumenting their urethra, and the pattern of inflammation differed to the expected
finding for auto-instrumentation. The urethral inflammation was a short length of circumferential ulceration in the bulbar urethra, rather than the localised injury on the posterior wall of the urethra just below the sphincter as expected from a self inflicted injury.

In conclusion, a series of seven pre-pubertal boys with bulbar urethritis of unknown aetiology is presented. Two of seven boys presented with urethral strictures prior to any intervention and two subsequently required dilatation. The aetiology of urethritis in these boys is possibly a variant of the auto-immune urethritis seen in Reiter's syndrome, may be due to an infection [perhaps Ureaplasma or Mycoplasma] and may be related to an immune deficiency. Boys presenting with this symptom complex should therefore have their immunological and HLA B27 status assessed, urethral swabs should be cultured for Chlamydia, Ureaplasma and Mycoplasma, and they should be endoscoped if they have a diminished urine stream or a prolonged period of symptoms. Effective therapy for this condition is not known, despite the fact that it is a relatively common presenting problem to a urological centre and, therefore, further study of the condition would help in understanding the disease.
OVERVIEW

The last chapter in this thesis has looked at a condition which produces narrowing in the bulbar urethra from an inflammatory process, rather than the congenital lesions discussed earlier. The principle reason for including urethritis in the study was to highlight the age range of the reports on which the original classification of both posterior and bulbar urethral anomalies were based. The effect of outflow obstruction on the subsequent bladder function is obviously important in management of any of the boys with significant obstruction with either a COPUM or a Cobb's collar, therefore this debate was also included. The case of Marion's disease was included to balance the dismissal of the Type II lesion suggested by Young et al. [1].

The principle reason for the compilation for these data was to better understand the clinical findings when compared to the common themes in the literature, with particular regard to the various terms found in previous reports. Those terms include posterior urethral diaphragm, posterior urethral valves, congenital stricture, Cobb's collar, Moormann's ring. The data from this study have identified two levels of obstruction, either at the posterior urethra or the bulbar urethra. The posterior urethral lesion is a membrane with a posterior hole with folds running onto it from the verumontanum, which when distorted looks to consist of valve leaflets (COPUM). Two significant associated findings were its position distal to the proximal extent of the external sphincter and the variable degree of obstruction produced. The congenital narrowing of the bulbar urethra was found to be either fibrous or muscular and is usually an incidental finding. The term Cobb's collar seems appropriate for this latter abnormality and it features are the common occurrence in boys with a syringocele and the separation from the verumontanum. This clarification of understanding of the male urethral congenital obstruction will hopefully stimulate further study and debate.
REFERENCES


33 Morgagni JB. Seats and causes of diseases investigated by anatomy; in five books, containing a great variety of dissections with remarks to which are added very accurate and copious indexes of the principal things and names therein contained. In 3rd ed. pp. 540-556. London, Millar,A. and Cadell T. Johnson and Payne, 1769.

34 Langenbeck JM. *Eine einfache und sichere methode des steinschnittes* Würzburg 1802.


45 Porak C. Valvule uretrale chez le fetus. Soc. Obstetricale, 1895.


53 Thompson GS. Note on a case of hydrenphrosis with urethral septum causing obstruction to urinary outflow. Lancet, 1907 1:506.


97 Earle Mr. Functions of the foetal kidney. *Lancet*, 1835 870.


122 Estes JM, MacGillivray TE, Hedrick MH, Adzick NS, Harrison MR. 

123 MacMahon RA, Renou PM, Shekelton PA, Paterson PJ. In-utero cystostomy. 

124 Hendren WH. Posterior urethral valves in boys. A broad clinical spectrum. *J.

125 Arnold SJ, Ginsberg A. Radiographic and photoendoscopic studies in posterior 

591.


128 Williams DI, Eckstein HB. Obstructive valves in the posterior urethra. *J.

129 Field PL, Stephens FD. Congenital urethral membranes causing urethral 

130 Crooks KK. The protean aspects of posterior urethral valves. *J. Urol.*, 1996 
126:763-766.


References - cont’d


146 ANZDATA. *Australia and New Zealand Transplant Registry* 1991.


Publications resulting from the study


Presentations resulting from the study


# Glossary

<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>ANZDATA</td>
<td>Australian and New Zealand Dialysis and Transplant Data Registry.</td>
</tr>
<tr>
<td>COPUM</td>
<td>Congenital Obstructive Posterior Urethral Membrane.</td>
</tr>
<tr>
<td>Cr</td>
<td>Creatinine.</td>
</tr>
<tr>
<td>GFR</td>
<td>Glomerular Filtration Rate.</td>
</tr>
<tr>
<td>GOS</td>
<td>Great Ormond Street, Hospital for Sick Children.</td>
</tr>
<tr>
<td>IVP</td>
<td>Intravenous Pyelogram.</td>
</tr>
<tr>
<td>MCU</td>
<td>Micturition Cystourethrogram.</td>
</tr>
<tr>
<td>US</td>
<td>Ultrasound.</td>
</tr>
</tbody>
</table>