AFRICAN JOURNAL OF MEDICINE and medical sciences

VOLUME 38 NUMBER 3 SEPTEMBER 2009

ISSN 1116—4077

Afr. J. Med. med. Sci. (2009) **38**, 221-225

Congenital genitourinary anomalies: an 8-year review in Benin City, Nigeria

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Summary

Congenital genitourinary anomalies are common indications for surgery in children. Identification of factors that may result in poor surgical outcome could be critical to policy formulation in developing countries. The records of children managed with congenital genitourinary anomalies at the University of Benin Teaching Hospital between January 1999 and December 2006 were retrospectively studied. They were aged between 10 days and 14 years (mean 4.3 ± 6.7 years) with a male/female ratio 18.8:1, and accounted for 801 (57.1%) of all children managed with congenital anomalies. Lower tract lesions 796 (99.4%) due mainly to hernia/hydrocele [483 (60.3%)] accounted for the majority of cases, but 105 (13.1%) lesions comprising of 71 hydrocele and 34 undescended testes resolved spontaneously during follow-up without surgical intervention. Of the 696 (86.9%) children operated, delay in presentation which allowed irreversible complications to develop affected outcomes. Therefore, of 26 children with posterior urethral valves, 9 deaths due to chronic renal failure were recorded, and this accounted for all the mortality in this study. Chronic inflammation in extrophy of the bladder resulted in bladder plate fibrosis which made them inelastic and difficult to close with resultant failed repair and persistent urinary incontinence. Children with undescended testis that presented before age 5 years had a better outcomes than those who presented later as atretic testes requiring orchidectomy were recorded after age 5 years. Overall, 600 (86.21%) children had successful surgeries, while 87 (12.5%) were with residual functional impairments.

Keywords: *Congenital, genitourinary, anomalies, 8-year, review.*

Résumé

Les anomalies génito-urinaires congénitales sont des indications communes pour la chirurgie chez les enfants. L'identification des facteurs qui peuvent entrainer les résultants médiocres de la chirurgie peuvent être critique dans la formulation des politiques dans les pays sous développés. Les registres des enfants traites ayant de telles anomalies au Centre Universitaire Hospitalier de Benin entre janvier 1999 a Décembre 2006 étaient analysées dans une étude rétrospective. Ils étaient âgés de 10 jours – 14 ans (moyenne d'âgé 4.3 ± 6.7ans) avec une proportion male/female de 18.8:1, et s estimait pour 801 (57.1%) de tous les enfants traites de ces anomalies. Les lésions inferieures [796 (99.4%)] du principalement a l'hernie/hydrocèle [483 (60.3%)] estimait dans la majorité des cas, mais 105 (13.1%) des lésions y comprise 71 des hydrocèles et 34 testicules suspendus et non-descendant était guérit durant la période de suivi sans intervention chirurgicale. Sur les 696 (86.9%) enfants opérés, la présentation tardive avec des complications irréversibles affectaient les résultats. Ainsi, sur les 26 enfants ayant eu une valve urétrale postérieure, 9 mouraient due a une complication rénale sévère et représentait les cas de mortalité de cette étude. L'inflammation chronique de la vessie résultait à une fibrose de la plaque de la vessie non-élastique et difficile à fermer entrainant un échec de réparation et une inconsistance urinaire persistante. Les enfants ayant leurs testicules suspendus dans l'abdomen traites a moins de 5 ans avaient des résultats satisfaisant que ceux qui se présentaient tardivement nécessitant une orchidectomie. En tout 600 (86.21%) d'enfants ont eu une chirurgie avec succès alors que 87 (12.5%) avaient un mauvais fonctionnement résiduel.

Introduction

Congenital genitourinary anomalies are very common and most lesions are obvious at birth. The common embryologic origin shared by genital and urinary systems makes embryologic derangements affect both systems. This results in a spectrum of congenital anomalies ranging from complete agenesis to ectopic location of organs in the systems [1]. During the last decade, particularly in developed countries, significant progress in the knowledge, including an improved understanding of epidemiology and pathophysiology of these anomalies, was made. The identification of etiologic genetic defects and significant improvements in surgical approaches led to a decrease in complications which resulted in improved cosmetic

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outcomes [2]. It is important to know and identify common congenital anomalies of external genitalia in order to determine the underlying genetic and/or endocrine disorders. Determination of the proper sex of an affected neonate at a very early stage of life is also necessary so as to raise him or her along the proper gender [3].

Appropriately timed antenatal ultrasound scan usually leads to prenatal diagnosis and makes it possible to treat obstructive and/or refluxing uropathies before the onset of clinical symptoms and irreversible damages. The prenatal diagnosis of hydronephrosis, unilateral renal agenesis, multicystic kidney diseases and posterior urethral valves results in initiation of intrauterine treatment or early postnatal investigations and treatment [4]. In developing countries, however, these are not possible because of lack of manpower and facilities needed for fetal diagnosis and intrauterine interventions. Even postnatal, delayed presentation is common in sub-Saharan Africa, particularly among the rural dwellers and the underprivileged. This also poses a great challenge because most of these patients present after irreversible complications have occurred [5-8]. An earlier study [3] in this subregion indicated that a large proportions of genitourinary anomalies are not diagnosed at birth and that outcomes of treatment are poor following post-natal repair unlike what obtains in advanced centres where results of intrauterine interventions and post natal repair are excellent [1,2,4]. The reasons for the poor outcomes of treatment in this subregion have, however, not been adequately highlighted. In view of this, it is important to audit and identify factors that influenced outcomes of treatment in our setting.

The aim of this eight-year retrospective study on children managed with congenital genitourinary abnormalities in our hospital is to determine the pattern of presentation, complications recorded, the surgical treatment offered and factors that influenced outcomes so as to make recommendations that may lead to improvements.

Materials and methods

An 8-year retrospective study based on children managed with congenital genitourinary anomalies at the University of Benin Teaching Hospital, Benin City, Edo State, Nigeria, between January 1999 and December 2006 was undertaken. Data extracted from the case files and collated included age, type of anomaly, organ affected, isolated/multiple lesions, clinical state on arrival, presence and type of complications, treatment options, intraoperative findings, post operative complications, factors that influenced outcomes of treatment and follow-up. The case files of children (n=21) which could not be retrieved for analyses were excluded.

Statistical analysis

The data obtained were analyzed using the statistical package for social sciences [SPSS version 11 (SPSS, Chicago, 111)] software. Continuous data were expressed as mean ± SD.

Results

Congenital genitourinary anomalies were very common indications for surgery during the period. There were children managed with congenital anomalies. Of these, a total of 696 (86.9%) had corrective surgeries while 105 (13.1%) were managed non-operatively (Table 1). They comprised of 659 males and 37 females with a male: female ratio 18.8:1, and were aged between 10 days and 14 years (mean 4.3± 6.7 years). Two children were seen with prune berry syndrome. Even though many lesions were obvious at birth, only 18 (2.6%) children presented during the neonatal period.

Lower genitourinary tract, with hernia and hydrocele together accounting for 483 (60.3%), were the most common anomalies. Twenty-eight of the children with hernias presented with incarceration and were managed on emergency bases. Of the 187 cases of hyrocele seen during the first six months of life, 71 underwent spontaneous resolution before 2 years during follow-up at the surgical out patient clinic. Excepting intrascrotal haematoma recorded in 13 children, surgical management was uneventful in those operated. Those with hypospadias accounted for 120 (15%) and presented at a relatively younger ages (mean age 2.1 ± 6.4 months) compared to others. Outcomes of repairs were initially very poor but this improved significantly during the study. On the whole, successful repairs were achieved in 86 (71.7%) cases while 34 (28.3%) had residual penile deformities and functional impairments. Children with undescended testes constituted 145 (18.1%) of the anomalies. Of the 63 cases diagnosed during the first three months of life, 34 descended before the first birth day and required no operation. Testicular volume was assessed with orchidometer (Prader's balls) during the period. Among the 111 children who had operation, 60 (54.1%) who presented before age five years had normal testicular volume. However, of the 51 (45.9%) who presented after five years, 10 (19.6%) had normal testicular volume, 28 (54.9%) had reduced testicular volume, while 13 (25.5%) had atretic testes. All the normal and reduced testicular

volumes had orchidopexy whereas the atretic testes necessitated orchidectomy.

Posterior urethral valves 26 (3.2%), bladder extrophy 10 (1.2%) and intersex disorders 9 (1.1%) were less commonly seen. Of the cases of posterior urethral valves, 9 presented after renal failure and sepsis had set in. They all died shortly after operation and accounted for the 1.1% overall mortality recorded during the period. Ten children, 5 males and 5 females, presented with bladder extrophy. Successful bladder closures were achieved in 6 children who were presented as neonates, but all had persistent urinary incontinence. Attempts at closure in the remaining 4 who presented after neonatal period failed. This was because bladder plate and soft tissues fibrosis as well as inelasticity of the pelvic bones had occurred. The four children subsequently defaulted from follow up. Congenital adrenal hyperplasia was the major cause of intersex disorders which resulted in anomalies of external genitalia. They were successfully repaired but vaginal stenosis requiring prolonged dilatation was recorded in two girls.

Epispadias 3 (0.4%), seen only among male children, cystic diseases of the kidney 3 (0.4%) and pelvi-ureteric junction obstruction 2 (0.3%) were very rare but repairs were successful without sequel. The 5 (0.6%) upper urinary anomalies seen during the period were unilateral with adequate compensation by the normal contralateral kidney. Overall, of the 801 children managed with congenital genitourinary anomalies, non-operative management was done and successful in 105 (13.1%), whereas of the 696 (86.9%) operated, 600 (86.2%) were successful, 87 (12.5%) had residual functional impairments while 9 (1.3%), all with posterior urethral valves, died as shown in the table.

Discussion

Congenital genitourinary anomalies are very common worldwide and an incidence of 1 per 1000 live birth has been reported in developed countries [1]. It is difficult to know the incidence in our setting due to poor data base but earlier surveys among children in Nigeria [5, 9,10] revealed a very high incidence. It accounted for 57.1% of all children with congenital anomalies in this study. Male preponderance with a male/female ratio 18.8:1 recorded in this series tallies with

Table: Congenital genitourinary anomalies managed in 8 years

Variables	Numbers	Percentage
Congenital anomalies in 8 years		
Congenital genitourinary anomalies	801	57.1
Other congenital anomalies	603	42.9
Total	1404	100
Management of genitourinary anomalies		
Operative management	696	86.9
Non-operative management	105	13.1
Total	801	100
Indications for operation		
Hernia/hydrocele	412	59.2
Hypospadias	120	17.3
Cryptorchidism	111	15.9
Posterior urethral valve	26	3.7
Bladder extrophy	10	1.4
Intersex disorders	9	1.3
Epispadias	3	0.4
Cystic diseases of the kidney	3	0.4
Pelvi-ureteric junction obstruction	2	0.3
Total	696	100
Outcomes of operative management		
Successful	600	86.2
Residual impairment	87	12.5
Deaths	9	1.3
Total	696	100

earlier reports [1,5-10]. This is because many common anomalies such as hernias, hydrocele, hypospadias, undescended testis and posterior urethral valves involved organs which are absent in females.

Although the majority of anomalies were obvious at birth, delayed presentation was common during the period. The mean age at presentation of all the anomalies in this series agrees with other reports [5-10] in this subregion but is at variance with report from developed countries [1-4] where intrauterine diagnoses and/or presentation within few hours of birth is the rule. Late presentation influenced outcomes of management especially among children with undescended testis, extrophy of the bladder and posterior urethral valves who presented after irreversible complications had occurred in a significant proportion of cases. Consequently, the mortality recorded was among children with posterior urethral valves who presented in renal failure and established sepsis which is similar to the experience of other authors [11]. Similarly, despite urinary incontinence, successful bladder closure was achieved among the six children with bladder extrophy who presented as neonates compared to failed attempt at closure recorded among those who presented much later, which agreed with similar studies [12-14]. Bladder and soft tissues fibrosis, inelastic bladder plate due to chronic inflammation, and rigidity of pelvic girdle resulted in failed repairs in this and other studies [12,13,15].

Also, children with undescended testes who presented within the first three months of birth had normal testicular volume with many undergoing spontaneous full scrotal descent before age 12 months. Whereas those who presented before age five years had normal testicular volume and all had orchidopexy, of those who presented after five years, 10 (19.6%) had normal testicular volume, 28 (54.9%) had reduced testicular volume, and 13 (25.5%) had atretic testes. Therefore, orchidectomy was inevitable in 25.5% of cases who presented after five years as similarly reported by previous authors [6-9]. Although hernia and hydrocele were the commonest anomalies as other authors also reported [5,16-18], pre- and post-operative morbidities were low. The 28 children who presented with incarceration and treated on emergency bases had uneventful post-operative courses.

Overall, congenital genitourinary anomalies affected mainly lower tracts in this study because only 0.6% patients presented with upper tracts involvements as earlier workers also reported [3,5,14]. Upper tract lesions were all unilateral with adequate compensation by the normal contralateral side. This partly explains why no morbidity or mortality was recorded despite the late presentation of these children. Until recently in this subregion, hypospadias repair was a major challenge with attendant poor outcomes [5,19]. This was due to poor knowledge of the pathology and lack of basic facilities required. However, recent advances significantly led to improved results in this and other centrers during the period [1,3,4].

Generally, outcomes of surgical management was poor in this setting because, of the 801 children, non-operative management was successful in 105 (13.1%), whereas of the 696 (86.9%) managed operatively, only 600 (86.2%) were successful, 87 (12.5%) had residual functional impairments and 9 (1.3%) deaths were recorded unlike a near hundred percent successful surgical management reported in developed centres [1,2,4]. Nevertheless, anaesthesia related complications were relatively low in this series compared to the experience of other authors in a similar setting [20,21]. This could be because the majority of children were operated after the neonatal period and many of the lesions were repaired using face mask/local anaesthesia. Follow-up of these patients, particularly those who required more than one surgery and those whose repair failed after first attempt, was a major problem in our setting which was not experienced by other authors in a more developed subregion [13].

In conclusion, congenital genitourinary anomaly was very common in our setting with majority of the lesions affecting the lower tracts. Outcomes of surgical managements were influenced by late presentation which allowed irreversible complications to be established on arrival. Therefore, chronic renal failure and sepsis accounted for the deaths among those with posterior urethral valves; bladder plate fibrosis resulted in failed repair among those with bladder extrophy and atretic testes led to many orchidectomies among those with undescended testes. It is hoped that public enlightenment campaign will result in early presentation, and improvement in prenatal/neonatal diagnoses as well as provision of basic surgical facilities will improve present results.

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Received: 25/10/07
Accepted: 06/07/09