Podium Session 4: Pediatric Urology
June 20, 2011, 1440-1540

POD-04.01

Bladder Neck Closure in Conjunction with Enterocystoplasty and Mitrofanoff Diversion for Complex Incontinence: Closing the Door for Good

Kavanagh Alexander, Afshar Kourosh, Scott Heidi, MacNeily Andrew
Department of Urololgical Sciences, The University of British Columbia, Vancouver, BC, Canada

Introduction: Bladder neck closure (BNC) is an irreversible procedure requiring compliance with catheterization of a cutaneous stoma and historically has been reserved for the definitive treatment of intractable incontinence after prior failed procedures. Recent literature suggests that BNC as a primary procedure does not result in extra morbidity and has a high success rate in compliant patients who are willing to be followed regularly. Our objective was to assess long-term outcomes of our series of patients undergoing BNC including: continence status; additional surgical interventions; post-operative complications; conception and sexual function; and satisfaction with BNC.

Methods: Institutional ethics approval was obtained to study all patients that have undergone BNC between 1990 and 2010 at the BC Children's Hospital. Patients were contacted via telephone interview by our nurse clinician.

Results: 25 consecutive patients [12 exstrophy, 13 neurogenic bladder (5 spina bifida, 4 cloacal anomaly, 2 spinal cord injury, 1 VATER and 1 UG sinus)] underwent BNC, augmentation cystoplasty and Mitrofanoff diversion. 20/25 patients (80%) had undergone 21 unsuccessful bladder neck surgeries prior to BNC. Median time from BNC was 63 mths (range 11-244). BNC was initially successful in 24/25 (96%) of patients with one patient requiring subsequent closure of a post-operative vesicovaginal fistula. Satisfaction rated strongly. 23/25 (92%) of Mitrofanoff diversions were continent for at least 4 hrs between CIC. 14 subsequent interventions were required in 9/25 patients (36%) including: cystolithotomy (12%), ESWL for upper tract stones (8%), Augment rupture (8%), injection of bulking agents for stomal leakage (8%), stomal revision for cutaneous stenosis/prolapse (8%), PCNL for stone (4%), open retrograde ureteral stenting for stone (4%) and transvesical revision of mitrofanoff stenosis (4%).

Conclusions: Bladder neck closure in conjunction with augmentation cystoplasty and Mitrofanoff diversion is an effective means of achieving continence in complex patients as a primary or secondary therapy. A significant minority of patients require subsequent intervention for complications arising from this surgery. Long term urological follow up into adulthood is essential.

POD-04.02

The Natural History of Severe Antenatal Hydronephrosis Due to Ureteropelvic Junction Syndrome

Riddell Jonathan[1], Jeldres Claudio[1], Lambert Raymond [2], Franc Guimond Julie[1], Barrieras Diego[1], Houle Anne Marie[1]
[1]Department of Pediatric Urology, CHU Sainte-Justine, Montreal, QC, Canada; [2]Department of Nuclear Medicine, CHU Sainte-Justine, Montreal, QC, Canada

Introduction: Severe antenatal hydronephrosis (AH) caused by ureteropelvic junction (UPJ) syndrome remains a complex management dilemma despite progress in medical imaging. Little is known about the natural history of untreated severe AH due to UPJ syndrome as most cases receive early intervention. The aim of this study is to present long-term data from our institution on expectant management of severe AH, to evaluate risk factors for future renal functional loss, and to validate our MAG3 Lasix drainage classification (Table 1).

Methods: From 1998 to 2008, cases from the CHU Sainte-Justine's renal scintigraphy database were reviewed , identifying 148 patients with SFU Grade 3 or 4 unilateral AH due to UPJ syndrome. 32 underwent initial pyeloplasty, 3 were lost to follow up, and 113 were observed with serial ultrasonography and MAG-3 studies . Initial postnatal ultrasound and renogram criteria , and our drainage classification , were evaluated in Cox linear regression models with >3% renal functional loss as the primary event.

Results: Of the 113 affected renal units, 82 (73%) were male, and 72(64%) leftsided. Over a median follow up of 55(4-180) months, 31 (27%) lost >3% function , and 19 (17%) had delayed pyeloplasty. Initial ultrasound criteria were not predictive of future renal functional loss. Decrease in SFU grade over the course of surveillance did predict maintenance of function (p<0.001, exp 0.545). Initial renogram criteria were not predictive of renal functional decline alone, but in combination in our classification system became highly prognostic of functional decline (Class 1: p=0.001 exp 1; Class 2 p=0.078 exp 2.5; Class 3 p=0.001 exp 26; Class4 p=0.001 exp 30).

Conclusion: Based on a median 5 year follow up, the majority of renal units with severe AH due to UPJ syndrome can safely be observed. Our classification system appears valid and clinically useful in terms of predicting the risk of future renal deterioration on the basis of the initial renographic evaluation. Affected renal units with Class 1 or 2 drainage and a stable ultrasound appearance can be safely observed in spite of severe hydronephrosis, while those with Class 3 and 4 drainage should be treated more aggressively.

Table 1. CHU Sainte-Justine's MAG3 Drainage Classification. POD-04.02

Class 1	Pre-Lasix drainage <70%; 20 min post-Lasix >40%
Class 2	20 min post-Lasix <40%; delayed (90minj 20min) drainage >50%
Class 3	20 min post-Lasix <40%; delayed (90minj 20min) drainage <50%
Class 4	Ipsilateral delay in cortical transit time >2min vs contralateral kidney

POD-04.03

Initial Experience with Laparoscopic Nephrectomy for Pediatric Uro-Oncology Cases

Romao Rodrigo[1], Weber Bryce[1], Figueroa Victor[1], Farhat Walid[1], Bagli Darius[1], Grant Ronald[2], Pippi Salle, Joao Luiz [1], Lorenzo Armando[1]
[1]Division of Urology, The Hospital for Sick Children, Toronto, ON, Canada; [2]Division of Hematology/ Oncology, The Hospital for Sick Children, Toronto, ON, Canada

Introduction: In adult urology laparoscopy has been increasingly adopted for the treatment of malignancies. In pediatrics there has been a slower trend towards the use of minimally invasive surgery for this purpose. Our aim is to present our initial experience with laparoscopic radical nephrectomy (LRN) for the treatment of malignant renal tumors in children.

Methods: Retrospective chart review of 6 children that underwent LRN and lymph node dissection for unilateral renal neoplasms over the past 3 years. Indication for laparoscopy was selective, based on clinical presentation, tumor size and family agreement.

Results: Median age at surgery was 9.5 years (2-17 years old). The tumor was left-sided in 4 and right-sided in 2 patients. Mean tumor size was 6.35 ± 1.13 cm. Only 1 patient received preoperative chemotherapy. Mode of presentation was blunt abdominal trauma in 2 patients, while incidental sonographic finding, gross hematuria, palpable abdominal mass and pain associated with tumor bleeding were the chief complaint in the remaining 4 patients. The operation was performed with 3 trocars (1x10 mm for camera, 2x5 mm for instruments). Lymph node sampling was performed in all and the adrenal gland was spared in 4 patients. There were no conversions. Three patients presented with ruptured tumors preoperatively and no ruptures were induced by the surgical procedure. Specimens were retrieved through a Pfannenstiel incision with an endoscopic specimen bag. Median operative time was 300 minutes (240-420). Median length of stay was 3 days (2-4) and all patients had a smooth postoperative recovery with straightforward pain management and excellent cosmetic result. Final pathological diagnosis was Wilms' tumor, renal cell carcinoma (RCC) with positive nodes and perivascular epithelioid cell tumor (PEComa) in 4, 1 and 1 patient, respectively. Surgical margins were negative for the non-ruptured tumors. Two patients underwent additional minimally invasive procedures, i.e. thoracoscopic resection of suspicious lung lesions that turned out not to be metastases. One patient developed an incisional hernia on the Pfannenstiel incision. At a median follow up of 4.5 months (1.5 to 25 months), all patients have been offered protocol-driven adjuvant treatment and have no evidence of recurrence.
Conclusions: LRN for malignant tumors in children appears feasible and safe in selected cases, with excellent surgical results. Long-term follow-up studies are needed to confirm comparable oncological outcomes to open surgery.

POD-04.04

Patterns of Presentation, Diagnosis and Gender Assignment in a Canadian Multidisciplinary Clinic of Disorders of Sex Development (DSD)

Romao Rodrigo[1], Bagli Darius[1], Lorenzo Armando[1], Farhat Walid[1], Chitayat David[2], Wherrett Diane[3], Allen Lisa[4], Zucker Ken[5], Neilson Barbara[6], Pippi Salle, Joao Luiz[1]
[1]Division of Urology, The Hospital for Sick Children, Toronto, ON, Canada; [2]Division of Clinical and Metabolic Genetics, The Hospital for Sick Children, Toronto, ON, Canada; [3]Division of Endocrinology, The Hospital for Sick Children, Toronto, ON, Canada; [4]Department of Paediatric and Adolescent Gynaecology, The Hospital for Sick Children, Toronto, ON, Canada; [5]Department of Psychiatry, The Hospital for Sick Children, Toronto, ON, Canada; [6]Department of Social Work, The Hospital for Sick Children, Toronto, ON, Canada

Introduction and Objectives: In 2006, a consensus statement was developed to standardize the nomenclature of DSD in an effort to facilitate communication among different specialists and abandon the concept of intersex. The authors wished to review their experience with these disorders in light of the newly proposed classification, highlighting the importance of multidisciplinary teamwork.
Methods: Retrospective chart review from 125 patients followed by a multidisciplinary DSD clinic in a 7-year period. Data collected included: karyotype, time of presentation (early × late), initial appearance of external genitalia and gonads evaluation, gender assignment and final diagnosis.
Results: Karyotype was XY in 60, XX in 51 and mosaic in 14 patients. Karyotype-based final diagnosis is depicted in Table 1. Overall, 109 patients (87%) presented as neonates or infants, whereas 16 (13%) were referred between 6 and 19 years of age (average 11,8).
XY Early presentation occurred in 87% of the patients, 58% had genital ambiguity in the form of undervirilization (proximal hypospadias with or without chordee) and 50% had at least one palpable gonad. In 21 patients (35%), there was no genital ambiguity (female appearance-14; male-7). Female gender assignment was 33%. Partial androgen insensitivity syndrome (PAIS) was the most common diagnosis (53%).
XX 84% of patients presented early. Clinical presentation was virilization of the external genitalia with no palpable gonads in virtually all patients. Congenital adrenal hyperplasia (CAH) was the final diagnosis in 84% and all patients in the XX group were raised as females except for 3 CAH late referrals.
Sex-chromosome DSD (Mosaic karyotype) 79% of patients had a 45XO/46XY karyotype. All presented early with genital ambiguity and were diagnosed with mixed gonadal dysgenesis (MGD). Gender assignment in this group was 8 (57%) female and 6 (43%) male.
Conclusions: Most patients were seen early in life with genital ambiguity. CAH and Partial Androgen Insensitivity Syndrome encompassed more than 50% of all diagnoses. Whilst gender assignment in XX DSD is usually straightforward, the same is not true for XY and Sex Chromosome DSD. Multidisciplinary team considerable deliberation aided in the decision-making process as well as family support, particularly for situations of discordant chromosomal and assigned gender.

POD-04.05

Outcome of a Prospective Randomized Trial Investigating Effect of Biofeedback on Self Esteem in Pediatric Patients with Dysfunctional Voiding

Weber Bryce[1], Pippi Salle, Joao Luiz[1], Buffett Angela[1], Guger Sharon[2], Leslie Bruno[1], Braga Luis[1], Bagli Darius[1], Farhat Walid[1], Lorenzo Armando[1]
[1]Division of Urology, Hospital for Sick Children, Toronto, ON, Canada; [2]Department of Psychology, Hospital for Sick Children, Toronto, ON, Canada

Introduction and Objectives: The objective of the present study was to evaluate the impact of biofeedback with validated, standardized questionnaires regarding perception of self-esteem as well as urinary symptoms in children referred with dysfunctional voiding. Children included in the study had a diagnosis of dysfunctional voiding, EMG activity during voiding phase of flow curve, and lower urinary tract symptoms.
Methods: Ninety seven patients were screened to enroll sixty patients which where then the distributed into two experimental groups via computer randomization: 1) the study group completed the questionnaires, history and flow rate prior to receiving biofeedback and received biofeedback the following week; 2) the control group completed the same questionnaires, history and flow rate at the initial visit and six weeks later prior to receiving biofeedback (delayed treatment group). All patients where then re-evaluated after six biofeedback sessions. Self perception domains were analyzed with the Self-Perception Profile for Children and Behavior Assessment Scale for Children questionnaires.
Results: No significant differences where seen in baseline characteristics (age, gender, history of UTI, reflux, constipation or anti-cholinergic therapy) or questionnaire scores. Patients undergoing biofeedback (study group) had significant and clinically relevant improvement (based on previous self-perception studies) and in total dysfunctional voiding scores (p=0.001), social acceptance (p=0.012), social stress (p=0.022), and self esteem (p=0.034).

Table 1. POD-04.04

	XY DSD	XX DSD	Sex-chromosome DSD	Total
CAH	4	43	2	49
PAIS	32	0	0	32
MGD	1	0	12	13
Complete Androgen Insensitivity Syndrome	10	0	0	10
OTHER	13	8	0	21
Total	60	51	14	125

Conclusions: This is the first randomized controlled study to show that biofeedback significantly improves self-esteem and social acceptance, while significantly decreasing social stress in children with dysfunctional voiding. The current results suggest that continued use of biofeedback has a positive impact on urinary function, which translates into improvement in self-perception.

POD-04.06

Do Infants with Low Grade Hydronephrosis and a History of Antenatally-Detected Hydronephrosis Benefit from Screening for Vesicoureteric Reflux?

Szymanski Konrad M, Al-Said A Nasser, Pippi Salle, Joao Luiz, Capolicchio John-Paul

Division of Urology, Montreal Children's Hospital, McGill University Health Center, Montreal, QC, Canada

Introduction and Objective: The natural history of vesicoureteric reflux (VUR) in children with antenatally detected low grade hydronephrosis (LGHN) is unknown. Screening infants with LGHN is unjustified if this type of VUR is benign. We present a cohort of patients with antenatally detected hydronephrosis (AHN), some of which were observed without a voiding cystourethrogram (VCUG) and compare outcomes to a classical group who were screened.

Methods: A cohort of 206 consecutive children with AHN (47% bilateral) presenting to Urology or Nephrology clinics were included in analysis. Cases with associated renal or bladder anomalies were excluded. Children with LGHN either underwent a screening VCUG or not, largely based on whether their care was managed by a Urologist or a Nephrologist. Cases of high-grade AHN (HGHN) routinely had VCUG. Primary outcome was a symptomatic or febrile urinary tract infection (UTI) confirmed with a catheterized urine culture. We used Mantel-Haenszel analysis to determine UTI risk factors over the first 2 years of follow-up.

Results: No UTIs were observed in patients with grade 1 hydronephrosis. UTIs in LGHN were only seen in the VCUG group (7 vs 0), including 1 post-VCUG UTI (14%). UTI incidence rate among children with LGHN was 2.93 UTIs/1000 patient-months (95% CI 0.76-5.10), while those with HGHN had 9.26 UTIs/1000 patient-months (95% CI 3.55-15.0) (p=0.02). This increased risk of UTI among HGHN persisted after correcting for gender and circumcision status (IRR=3.17, 95% CI 1.20-8.41, p=0.01) and was marginally significant after additionally correcting for VUR status (IRR=2.48, 95% CI 0.96-6.44, p=0.053). VUR, gender and circumcision were not independent UTI risk factors ($p \geq 0.21$). Among children with LGHN who had a VCUG (66%), VUR was detected in 9.2% vs 14% in HGHN (p=0.42).

Conclusions: Our data suggests that children with LGHN and otherwise normal kidneys and bladder do not benefit from VCUG screening. A prospective randomized study is required to confirm the long-term safety of this approach. HGHN appears to be a 3-fold risk factor for UTI.