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Prognosis for children born with multicystic dysplastic kidneys

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Objectives: To review our experience with multicystic dysplastic kidneys (MCDK) comparing nephrectomy with careful follow-up, to plan for the future management of this disorder.
Patients and methods: A retrospective study was made of 26 children with MCDK who underwent nephrectomy between 1985 and 1995, and 11 children born between 1993 and 1994 who were followed conservatively for at least one year.
Results: The 26 nephrectomized children were aged between 8 days and 10·5 years (median age 7·5 months); 20 were diagnosed pre-natally. Seven of 26 (27%) had contralateral pathology (three contralateral reflux and four PUJ obstruction). Indications for surgery in the pre-natally diagnosed group were: increasing size (7) with a median follow-up of 9 months, unchanged size (7), large MCDK (2), concomitant contralateral surgery (2), solid lesion (1) and hypertension (1). Among 16 children born 1993–94, only five underwent nephrectomy; the indications were increasing size (3), large size (1) and unchanged (1). Among the remaining 11 children, the size decreased in eight (after 1 month to 2 years), increased in two and was unchanged in one.
Conclusion: These findings suggest that more than half of the MCDKs decrease in size over 1–2 years, but completeness of eventual involution remains in doubt. A substantial number of MCDKs increased in size during the same time (19–35%). Prolonged follow-up must be weighed against today's very safe nephrectomy. We suggest nephrectomy if the MCDK increases in size or fails to show substantial involution during the first 2 years.

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Vesico-ureteric reflux and dysfunctional voiders: resolution rate and age at diagnosis

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Introduction: Children with VUR frequently exhibit signs of dysfunctional voiding (DV) and VUR often resolves when bladder training or maturation corrects DV, suggesting that high pressures with DV contribute to VUR. We studied VUR with and without DV with age at diagnosis and rates of resolution.
Patients and methods: Between 1981–1984 222 patients with VUR were seen; 59 had DV (wetting, urgency, hesitancy; posturing). Resolution rates were estimated by the Kaplan–Meier method. Log rank and Cox regression comparisons were made for age at diagnosis and resolution of VUR.
Results: The mean follow-up was 5·9 years, the age at diagnosis of VUR for normal voiding was 3·9 years and for DV 4·9 years ($P = 0·003$). Overall, resolution of VUR was similar in both groups, but was slower with DV before the age of 9 years and more rapid after 9 years than for normal voiding ($P < 0·05$).
Conclusion: VUR in children with DV presents later than in children with normal voiding. More rapid resolution can be anticipated in this group after the age of 9 years, perhaps as the result of lower voiding pressures with maturation or bladder training with correction of DV. Urologists should be cautious in recommending early surgery to correct VUR associated with DV as not only may surgery prove to be unnecessary, it also carries a greater complication rate.

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One-stage orchidopexy for high undescended testes using low testicular vascular ligation: an alternative to the Fowler–Stephens technique

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Introduction: Since popularization in 1959, the Fowler–Stephens one-stage orchidopexy, involving ligation of the testicular vessels high above the testis, has been the standard for the treatment of high undescended testes. Based on recent studies of testicular vascular anatomy, we hypothesized that vascular ligation is best performed adjacent to the testis.
Patients and methods: During the past 5 years, we performed one-stage orchidopexy for high undescended testis through a skin-crease inguinal incision by ligating the spermatic vessels adjacent to the testis, and dividing the peritoneum between the testis and vas in 33 patients with 37 high undescended testes located intra-abdominally or near the internal inguinal ring.
Results: The one-year follow-up showed a viable testis by palpation and/or Doppler flow study in 91%.
Conclusion: These results are comparable or better than standard one-stage or laparoscopic two-stage Fowler–Stephens orchidopexy, and suggest that in the treatment of high undescended testes (1) high ligation is not necessary for testis viability, (2) vascular anatomy supports low ligation, (3) low ligation may enhance testis viability by reducing tension and improving collateral flow, (4) low ligation simplifies the surgical technique so that an inguinal incision can be used in all cases, and (5) deliberate two-stage orchidopexy is difficult to justify.

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Management of the intraperitoneal testis

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Introduction: The two most controversial questions in the management of the intraperitoneal testis: Is the intraperitoneal testis worth bringing into the scrotum and what is the best method to do so?
Patients and methods: Forty-four consecutive intraperitoneal testes were managed. All underwent a transperitoneal orchidopexy and, if vessel length did not permit a successful operation, an autotransplantation was done at the same operation. A tubular spermatic index and tubular diameter was obtained on each testis using plastic-embedded sections and toluidine-blue staining. The mean age of the child undergoing transperitoneal orchidopexy was 23 months and the mean operative time was 2·5h, while that of the child undergoing autotransplantation was 16·6 months with an operative time of 6·8h. Hospital stay was 1·5 days and 3 days, the shorter stay for the transperitoneal orchidopexy.
Results: Thirty-eight (86%) of the testes were brought into the scrotum with a transperitoneal orchidopexy and of these, 32 (84%) were successful (testicle of good size and consistency 3 months post-operatively). Six (14%) were managed by autotransplantation and of these four were successful. Two underwent a successful second operation when the testicle retracted upward and both were successful. The overall success with one operation was 77% and, including the two with a second operation, 86% of the total group had a successful outcome. The tubular spermatic indices of the intraperitoneal testes after one year or less was 0·493, after 1 to 2 years 0·475 and 0·54 after 2–4 years. These were comparable to intracanalicular testes of the same age.
Conclusions: The intraperitoneal testis is a good quality testis in the first few years of life and can be successfully managed with one operation with a success rate of 77%.

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Spontaneous rupture of augmentation entero-cystoplasty in children

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Introduction: To review the diagnosis, treatment and outcome of spontaneous perforations in augmented bladders in the paediatric population.

Patients and methods: Between 1980 and 1994, 250 augmentation cystoplasties were performed at this institution. Ten patients had 14 perforations. Eight had bladder exstrophy and two had neuropathic bladders (myelomeningocele in one and tethered cord in the other). There were eight males, aged 4–16 years (mean 7·3 years) and two females aged 4 and 16 years at the time of augmentation. One male patient with a neuropathic bladder and detubularized sigmoid colon cystoplasty had two perforations at 3 and 4 years after augmentation. One female patient with bladder exstrophy and detubularized ileo-cystoplasty and bladder neck reconstruction had four perforations at 7 months, 11 months, 2 years and 6 years after surgery. The interval between augmentation and perforation ranged from 7 months to 11 years (mean 4·8 years). Ileum was used in six and sigmoid colon was used in four patients. All patients with exstrophy had increased outlet resistance with a Young-Dees reconstruction (8), a Kropp onlay procedure (1), and an artificial sphincter (1). All 10 were on clean intermittent catheterization.
Results: Abdominal pain was present in all cases, with obvious peritonitis and pyrexia in eight. Cystography was performed in five cases, but showed urinary extravasation in only one. Two patients presented with a clear history of trauma. The time from the last catheterization to perforation ranged from 3–9 h (mean 6). Laparotomy and repair of the perforation was carried out in all cases, followed by catheter drainage for 2 weeks. All perforations were in the enterocystoplasty segments. There was no long-term morbidity or mortality.
Conclusion: Non-compliance in performing regular catheterization, presumably causing over-distension of the augmented bladder, appeared to be the precipitating event leading to perforation. Prompt diagnosis based on a high degree of clinical suspicion followed by laparotomy and repair minimized morbidity and mortality in this potentially lethal condition.

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Combined use of Mitrofanoff channel and ACE procedure for urinary and faecal incontinence

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Introduction: Faecal soiling occurs frequently in association with urinary wetting in children undergoing major reconstructive urological operations. To treat double incontinence, we constructed a Mitrofanoff conduit and a channel for antegrade continence enemas (ACE) in 16 patients between 1991 and 1995.
Patients and methods: The underlying abnormalities included mainly spinal lesions and bladder exstrophy, usually complicated with sacral agenesis, imperforate anus and cloacal variants. Patients were aged 2–13 (mean 7·8) years. In 11 patients, both procedures were performed simultaneously. The appendix was used to construct an ACE channel in seven cases, Mitrofanoff in five and it was long enough to be divided and used for both in one. Unfortunately, the appendix was missing or unsuitable in three cases. The alternative conduits used for ACE were a caecal flap (7) and ileum (1) while the ureter (4), ileum (5) and detrusor tube (1) were used to establish Mitrofanoff channels. The stomas were constructed either according to the V-flap or *VZQ* techniques and situated in most patients (12) in close proximity, in the right lower abdominal quadrant.
Results: Post-operative recovery was generally uneventful, except for one patient with abscess formation near the ACE stoma. Nine patients needed dilatations or minor revisions due to difficulties in catheterizations of the ACE (4), Mitrofanoff (3) or both (2). Subsequently, three patients underwent operations for reconstruction of two ACE channels (caecal flap and ileum) and one detrusor tube. At present, 14 patients are dry on regular CISC using 10–12 F catheters. The outcome of the ACE channels was satisfactory in 13 patients who are clean or rarely soil. The failures include one case of severe constipation necessitating colostomy and two other patients (one non-compliant patient who stopped catheterizing regularly) whose channels subsequently closed.
Conclusion: Synchronous construction of ACE and Mitrofanoff channels was successful in most doubly incontinent patients. Selection of patients with high motivation is important in obtaining satisfactory results.

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The anatomy and reconstruction of the adult female genitalia in classical bladder exstrophy

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Introduction: This paper addresses the anatomical anomaly of the external genitalia in females with bladder exstrophy. Through experience of a large series of patients, we sought to optimize both function and cosmesis in reconstruction of the external genitalia.
Patients and methods: Details of genital reconstruction, sexual function and obstetric history in 47 female patients with bladder exstrophy followed in an adolescent clinic were reviewed. The anatomy of each patient was studied prospectively. The principles of reconstruction include opening of the introitus (vaginoplasty), fusion of the labia anteriorly, and reconstruction of the mons pubis (vulvoplasty). Introital stenosis of any significant degree requires reconstruction to allow penetrative intercourse. Vulvoplasty can be combined with vaginoplasty to achieve satisfactory cosmesis.
Results: (1) Genital anatomy: the vagina in female exstrophy is foreshortened, horizontal, anteriorly displaced and stenosed. The fourchette is deformed by failure of fusion of the clitoris and anterior labia. The mons is also widely spaced and the hairline is separated by scar tissue.
(2) Reconstruction: 27 of the 47 patients have had vaginoplasty, and eight of these also had vulvoplasty. All 27 reconstructed patients have been sexually active. Only six of the 20 non-reconstructed patients are desirous of and able to have a form of penetrative intercourse. There have been 20 pregnancies in 13 patients. Five were delivered per vaginum and eight by caesarean section.
Conclusion: Females with bladder exstrophy have grossly abnormal external genitalia. Reconstruction is required for sexual function as normal penetrative intercourse is virtually impossible without reconstruction. Vulvoplasty improves the otherwise abnormal appearance.

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Comparison of various methods of repairing the symphysis pubis in bladder exstrophy by tensile testing

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Introduction: The properties of the immature bone, ligaments and cartilage of the symphysis in newborns with bladder exstrophy are unknown and it is not clear what methods of fixation can best hold these structures together. We compared various methods of fixation by using different techniques to fix the symphysis pubis in an immature animal pelvis model and then to compare their biomechanical properties and investigate the modes of failure for each repair.
Materials and methods: The pelves of piglets (body weight 3·5–5·9 kg) were weighed and dissected. The symphysis was left intact in the control group and then in groups 2–9, the symphysis pubis was cut completely and repaired using one of eight different techniques. Each pelvis was then placed in a specially designed jig for tensile tensing on an MTS machine. Tensile force was applied to each pelvis and the mean load failure and stiffness for each repair technique calculated.
Results: Of all the repair techniques used, a horizontal mattress nylon suture was able to withstand a higher load-to-failure force. The group of pelves repaired with Mersilene tape proved to be the stiffest when loads were applied, compared to other suturing techniques.
Conclusion: The pelvic bones of the piglet are partially ossified, as in the newborn infant, with a significant amount of cartilage still present in the symphysis. While all repair techniques are weak compared to an intact symphysis, the best load-to-failure closure was achieved with a Number 2 nylon stitch, although it was still less than that of intact bone. The best stiffness achieved with Mersilene tape was still less than one-half that for an intact symphysis. Future investigation is warranted to find methods which better approximate the strength of an intact pubic symphysis in the newborn with bladder exstrophy.