

PEDIATRIC UROLOGY _____

A preliminary investigation into quality of life, psychological distress and social competence in children with cloacal exstrophy

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Purpose: Cloacal exstrophy is a complex multisystem anomaly. Due to ambiguous genitalia gender assignment or reassignment is common. The psychological, emotional and behavioral impact of this condition has rarely been investigated.

Materials and Methods: We recruited 8 children with cloacal exstrophy born with genital ambiguity and a control group of 12 with cloacal anomalies born without genital ambiguity were recruited via urology-endocrine clinics at Great Ormond Street Hospital for Children. Patient age was 5 to 18 years (average 11.3). The child behavior checklist, child health related quality of life and social cognition questionnaire were administered to assess perceived levels of social competence and adjustment, emotional and behavioral distress, and perceived quality of life.

Results: Social and behavioral competence as well as psychological problems were comparable with normative data for the 2 groups. There were no statistically significant differences in the 2 groups on any competence, problem or social adjustment scale. A quality of life measure again revealed no significant differences in the groups. The scores obtained were comparable with those reported for other chronic illnesses.

Conclusions: Results suggest that being born with cloacal exstrophy or anomaly and gender assignment or reassignment does not necessarily result in childhood psychological, emotional or behavioral distress and/or problems, lower levels of social competence or subjective reports of poor quality of life. It is suggested that longitudinal and larger studies are required to assess the long-term implications of this condition.

Editorial Comment

This is an extremely important report of the psychological evaluation of children with cloacal exstrophy, with particular emphasis on 6 patients with 46 XY chromosomes who were raised as females. The researcher was blinded as to the chromosomal diagnosis in the cases and there was a control group of patients with cloacal anomalies, but not cloacal exstrophy or gender conversion. The authors found no statistical differences between the 2 groups, with scores comparable to those reported for other chronic illnesses.

The gender of rearing is highly controversial in patients with cloacal exstrophy and 46 XY chromosomes. Because the phallic structures are usually too small to be adequately reconstructed as a normal male phallus, most patients have, in the past, been raised as females. On the other hand, they have already had their central nervous system “imprinted” with androgens prenatally. Recently, fears of gender dysphoria and ultimately reassignment (no good published reports in peer-reviewed journals), have led many physicians to recommend raising these children as males, despite the fact that they will have an inadequate phallus. Hence the development of these patients as normal females is of major interest. On the other hand, this study has several cautions. First, follow-up is still too short with a mean age of 11.6 years. Second, 5 of the 16 patients eligible refused to be interviewed and 3 of the others were undergoing psychiatric treatment and were deemed ineligible. Data from these patients and follow-up through puberty could ultimately change the results. We anxiously await more information in this critical area.

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Natural history of neonatal reflux associated with prenatal hydronephrosis: long-term results of a prospective study

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Purpose: We have previously reported on patients with neonatal vesicoureteral reflux followed conservatively. The current study is a long-term follow-up of our prospective expectant management protocol for the overall cohort.

Materials and Methods: Between 1993 and 1998, 31 of 260 patients with prenatal hydronephrosis had vesicoureteral reflux and were followed prospectively. Outcome analysis was done on 25 patients, excluding 6 who underwent surgery, with the end point of complete resolution or improvement of reflux using our previously reported Kaplan-Meier survival curve, urinary tract infection, dysfunctional voiding, and changes in renal function or growth, somatic growth and hypertension.

Results: Of the 25 cases reflux was grades I to V in 7%, 20%, 34%, 16% and 23%, respectively. Reflux resolved in 13 patients (52%) and improved in 6 (24%). Grades I to V disease resolved in 100%, 77%, 53%, 28% and 40% of refluxing units, respectively. The improvement rate for grades III to V reflux was 13%, 14% and 30%, respectively. Breakthrough urinary tract infection occurred in 4 patients with grades IV and V reflux, and dysfunctional voiding developed in 5. Follow-up renal scans showed 19% and 17% decreased differential function in 2 units without new scars. There was no difference in renal length in patients with resolved versus persistent reflux or low versus high grade reflux. All patients had normal somatic growth at the 4-year follow-up and none had hypertension.

Conclusions: Expectant management was effective in the majority of cases and associated with a low urinary tract infection rate. Neonatal vesicoureteral reflux resolved or improved in 76% of our patients by age 4 years without somatic growth retardation or hypertension. High grade reflux resolved or improved in 59% of the units and showed normal renal growth with expectant management.

Editorial Comment

This is an interesting review of the mid-term follow-up (median of 44 months) of children diagnosed with vesicoureteral reflux as neonates. The reflux resolved spontaneously in 52%. Four patients had UTIs and dysfunctional voiding developed in 5. Two patients developed decreased renal function. The authors conclude that expectant management was effective in the majority of cases.

This paper is an interesting contribution, but the interpretation of the data seems skewed and the conclusions are arguable. In the first instance, 6 patients underwent surgery and are excluded. In general, studies are better reported in an "intent-to-treat" format. Excluding the 6 patients needing surgery changes the results. Furthermore, not all patients underwent follow-up DMSA scans, hence there may have been more than 2 cases of renal injury. Finally, despite 4 years of antimicrobial therapy, 48% of patients did not have resolution of their reflux. My personal opinion is that this study could also be used to argue more forcefully for more surgical therapy.

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