Correspondence

Archives of Disease in Childhood, 1976, **51**, 324.

Cryptorchidism: is routine intravenous pyelography indicated?

Sir,

Increased incidence of upper urinary tract malformations has been reported in cryptorchids (Felton, 1959; Grossman and Ririe, 1968; Farrington and Kerr, 1969; Donohue, Utley, and Maling, 1973; Watson, Lennox, and Gangai, 1974; Engel *et al.*, 1975). Routine intravenous pyelography (IVP) has therefore been recommended. Recently, Watson *et al.* (1974) claimed that this procedure was of limited value since few disorders of clinical significance were discovered on IVP of cryptorchids. For further elucidation of this question IVP was performed in 166 cryptorchid boys aged 2.5–16.8 years. Retention was unilateral in 96 and bilateral in 47 patients. 15 boys had unilateral and 7 bilateral anorchia. Episodes of pyuria in the clinical histories were especially noted. A detailed clinical description of the patients will be given elsewhere (Waalder, 1976).

The Table shows the results of IVP in relation to episodes of pyuria. A normal pyelogram was found in 93% of the boys. Minor congenital abnormalities were discovered in 10 boys. 2 of these boys had bilateral and 7 unilateral undescended testes whereas one had unilateral anorchia. One boy with bilateral testicular retention had crossed renal ectopia with moderate hydronephrosis and variable cortical thickness. He had been treated for febrile pyuria in infancy. Unilateral chronic pyelonephritic shrinkage was discovered in a 13-year-old boy with mild hypertension. This renal defect was regarded as an acquired disorder. Only 6 of the patients had had episodes of pyuria and in 3 of them the IVP was normal.

The incidence of minor upper urinary tract anomalies in the general population is difficult to assess. Renal malrotation, forked ureters, and pelviureteric junction stenosis are usually regarded as frequent phenomena. In the necropsy series of Felton (1959) 2% of 152 boys had unsuspected abnormalities of the upper urinary tract. Nordmark (1948) investigated the pyelograms of 4774 patients and found ureteral duplications in 2.8%.

In this study only 3.6% of the boys had had episodes of pyuria and 6.6% had minor or major anomalies of the upper urinary tract discovered on routine IVP. According to other reports (referred to above) urinary tract anomalies were discovered on IVP in 12–26% of cryptorchids. Watson *et al.* (1974) reviewed the results of 400 IVPs taken in asymptomatic cryptorchids; in 10% minor anomalies were found, and major abnormalities in 3%. The variations of reported incidences may partly be caused by differences in the selection of patients. In our study only 2 patients (1.2%) had significant disorders discovered by IVP. In both these boys there was an additional indication for examination (pyuria in one, mild hypertension in the other). We agree with the conclusion of Watson *et al.* (1974) that routine IVP of cryptorchids will give a low yield of significant upper tract disease. However, any additional suspicion of such disease or the combination of cryptorchidism with other congenital anomalies is an indication for IVP.

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TABLE

Intravenous pyelography in 166 cryptorchid boys with or without a history of pyuria

Result of intravenous pyelography	Without episode(s) of pyuria	With episode(s) of pyuria	Total (%)
Normal	151	3	154 (92.8)
Minor anomalies			} 10 (6.0)
Unilateral duplicated pelvis with forked ureter	2	2	
Unilateral slight pelviureteric obstruction	5	0	
Bilateral slight renal malrotation	1	0	
Crossed renal ectopia	0	1	1 (0.6)
Unilateral chronic pyelonephritic shrinkage	1	0	1 (0.6)
Total (%)	160 (96.4)	6 (3.6)	166 (100)

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Genetic miscounselling in muscular dystrophy

Sir,

Raised serum creatine phosphokinase (CPK) is now widely used in the diagnosis of Duchenne muscular dystrophy and in detection of female carriers. While a significant rise in CPK in the potential carrier is presumptive confirmation of her carrier status, a normal result does not exclude it. About 20–30% of genetically definite carriers have consistently normal enzyme levels. In addition, the level of CPK may fluctuate in the same individual and it is advisable to repeat the examination on at least three occasions. In possible carriers with a normal CPK level additional help in confirmation of carrier status may be obtained from overt abnormality on quantitative electromyography or muscle biopsy (for review see Dubowitz, 1975).

Case report

A 19-year-old female with 2 brothers who had died of classical Duchenne muscular dystrophy at the ages of 22 and 20 years, respectively, sought genetic counselling at her local hospital at the time of her marriage. On the basis of a single normal serum CPK result she was advised that she was not a carrier and could safely have children. After a period of prolonged infertility she became pregnant 7 years later after clomiphene stimulation and artificial insemination and delivered a normal male infant at term.

A cord blood sample for CPK was not obtained but a venous sample from the infant on the 3rd day showed a level >1000 IU/l; at the age of 6 weeks two samples were 5000 and 4900 IU/l, respectively; and at 7 weeks >1000 IU/l. The mother's levels at 4 and 5 weeks post partum were 95 and 60 IU/l. (Method used: Boehringer kit; normal for adults 0–130 IU/l.) At the age of 11 weeks the infant's CPK was 1090 IU/l, and that of the mother 22 IU/l. (Method: Boehringer (25°C), normal for adult female <60 IU/l.) Electromyography

on the infant was normal but a muscle biopsy from the rectus femoris showed unequivocal changes of early dystrophy.

This case illustrates the difficulty of excluding Duchenne dystrophy in a possible female carrier with normal CPK levels. As she already had 2 affected brothers one can assume that her mother was a definite carrier and she thus already had potentially a 50% chance of being a carrier on genetic grounds. In such instances it is advisable for patients to be referred to special centres for more detailed assessment of their carrier status.

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Insulin response to glucagon in short children

Sir,

Dr. Karp and his colleagues have reported low insulin responses to glucose and arginine stimulation in thin children with familial short stature and have recently shown that glucagon injection produces a normal rise in insulin in these patients (Karp, Laron, and Doron, 1975). It is possible that the poor insulin response to glucose in these patients may be secondary to their growth failure, rather than an aetiological factor. There is evidence that serum insulin levels after oral glucose vary with age and low levels are common in small children (Grant, 1968; Paulsen, 1973; Rosenbloom *et al.*, 1975). The explanation for this is not known but it is possible that sensitivity to endogenous insulin falls with increasing maturity. The low insulin responses to glucose which were found by Karp *et al.* in their patients may be related to physical 'immaturity', and it would be most interesting to learn whether the results were still abnormal when compared with those obtained in normal children of comparable size and skeletal maturity.

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