ASSOCIATION OF CONGENITAL RENAL AND INTESTINAL LESIONS

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TRACHEO-OESOPHAGEAL fistula and oesophageal atresia (TOF) duodenal atresia, anorectal anomaly and exomphalos are known to have a significant association with renal abnormalities.[1-4] The incidence of the association varies depending on the primary gastrointestinal condition and the reported series but is sufficiently common in all to merit consideration of the routine investigation of neonates with these conditions by way of intravenous pyelogram and micturating cystogram. Such a policy had gradually been introduced since 1977 in the Royal Belfast Hospital for Sick Children and the diagnostic yield of this policy to December 1982 is presented.

METHODS

The records of all neonates with primary diagnosis of tracheo-oesophageal fistula, duodenal atresia, imperforate anus or exomphalos were investigated to determine the presence or absence of renal abnormalities. In cases where the child was deceased and no renal study was performed, post mortem records were examined.

The policy to investigate these children was not uniformly applied initially but has been almost total since 1980.

RESULTS

Eighty-three cases were examined, 31 had no radiological investigation of the urinary tract, 9 had an incomplete investigation i.e. only one of either intravenous pyelogram or micturating cystogram. Thirty-seven abnormalities were discovered in 28 patients (54.6 per cent of the cases with partial or complete radiological investigation or 65.1 per cent of the cases with full radiological investigation). Fourteen cases died before investigation: one abnormality was reported at post mortem in this group—ureteral obstruction queried as secondary to posterior urethral valves.

Of the 42 T.O.F. examined eight cases not radiologically investigated developed urinary tract infections which would now be regarded as an indication for intravenous pyelogram and micturating cystogram in itself. In none of these cases was radiological investigation carried out. The reasons for these omissions could not be ascertained.

The 14 cases with duodenal atresia contained eight with Down's syndrome. Six died before investigation (4 Down's syndrome). No abnormalities were revealed at post mortem.

Six cases out of 25 cases of imperforate anus died before investigation. No abnormalities were revealed at post mortem. Thirteen cases had abnormalities other than an intestinal fistula to the urinary tract.

The occurrence of normal and abnormal urinary tracts in the various groups is presented in Table 1.

TABLE 1

Results of Clinical and Radiological Investigations

Diagnosis	*Abnormal*	*Normal*	*Incomplete*	*Early Deaths*	*Total*
Oesophageal atresia tracheo-oesophageal fistula	12	13	17	—	42
Duodenal atresia	2	1	5	6	14
Anorectal anomalies	15	4	—	6	25
Exomphalos	—	3	—	6	9

Table 2 shows the distribution of more common urinary anomalies in the series. Other less common disorders included crossed renal ectopia (4 cases), persistent urachus (1 case), urogenital sinus (1 case), vesico-ureteric junction obstruction (1 case): renal dysplasia (2 renal moieties) in two separate cases.

TABLE 2

Distribution of Most Common Anomalies

Diagnosis (single moieties)	*Tracheo-oesophageal Fistula (cases)*	*Anorectal Anomalies (cases)*	*Duodenal Atresia (cases)*
Vesico-reflux —22	8	7	2
Agenesis — 4	3	1	—
Hydronephrosis — 5	2	2	—

DISCUSSION

The co-existence of multiple congenital abnormalities in many situations is fully documented and it is consequently recognised that certain associations exist in specific circumstances. Such a situation is the case with four major conditions affecting the gastrointestinal tract, T.O.F., duodenal atresia, imperforate anus and exomphalos all of which carry a high association with cardiac, skeletal and renal abnormalities.

It is with the renal lesion in these conditions that we are concerned in this study as they occur sufficiently frequently[1-4] to merit routine investigation following diagnosis of the gastrointestinal lesion, and cover the entire spectrum of renal abnormalities making prediction of the nature of the anomaly impossible except in the anorectal anomalies where a fistula from the terminal bowel to the urinary tract is assumed if it is not present in the genital tract or perineum.[4] It is therefore justifiable to investigate these children radiologically. Having pursued an

investigative policy for six years, which was initially not enforced consistently but now has been routinely applied for two years, our findings underline that this policy should continue as 50 per cent of the cases investigated had abnormalities which were significant. Although the cases of exomphalos did not reveal any abnormalities of the renal tract it is assumed that this is accounted for by the small number and the high percentage of deaths prior to investigation: on the basis of the experience of other authors[3] we will continue to investigate exomphalos for associated renal anomalies.

It may be argued that if these cases are of long term significance—this particularly applied to vesico-ureteric reflux which may resolve spontaneously — they would eventually become manifest clinically and that pursuing an expectant policy would obviate investigation of those with no abnormality. The danger with such an approach is identical to that with urinary tract infection in children generally, namely that the symptomatology is notoriously misleading and often minor even in the presence of gross pathology. There is therefore a great risk in what is already a threatened group of patients that a correctable lesion may be overlooked until organ failure is unavoidable.

SUMMARY

Eighty-three neonates with congenital gastrointestinal disorders carrying a known association with urinary tract abnormalities were investigated. Thirty-seven abnormalities were discovered in 28 patients.

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