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Neonatal duodenal obstruction with emphasis on cases with Down's syndrome

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SUMMARY

In the past 31 years, 47 cases of duodenal obstruction were admitted to the Royal Belfast Hospital for Sick Children. Thirty-six per cent of these had Down's syndrome. The crude mortality was 48% but this figure was reduced to 33% in the last five years of the study. The combination of duodenal obstruction, Down's syndrome and any other abnormality carried an 89% risk of mortality.

INTRODUCTION

Duodenal atresia occurs in approximately 1/1,500 live births. Treatment of neonatal duodenal obstruction in isolation should have a successful outcome. However, this remains a dangerous condition because of the association with abnormalities in other systems, particularly cardiac and renal. The association with Down's syndrome produces the major difficulty both pre- and post-natally. This review was therefore undertaken to establish the relative prognosis of Down's syndrome with duodenal atresia.

METHODS

The hospital records for the period 1954-1985 inclusive were examined and the following details documented for each period: gestational age, birth weight, date of birth, sex, presence of Down's syndrome and other congenital abnormalities, condition on admission, details of surgery and any subsequent complication.

RESULTS

The period under study was 1954-85 inclusive. There were 47 cases recorded, 24 boys and 23 girls. Down's syndrome was present in 17 cases (36%) — 12 boys and five girls. The crude mortality for the group as a whole was 47%, and 65% for the Down's syndrome sub-group. In the last five years of the study, five of the 15 cases (33%) did not survive. The mean gestational age was 36.5 weeks for non-Down's cases, and 37 weeks for Down's, whilst the mean birth weight was 2.58 kg for both groups. All the survivors under 2.5 kg and less than 34 weeks' gestation were born in the last 10 years. Table I shows the frequency and mortality with a subdivision for Down's syndrome over two 10-year and one

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TABLE I

Duodenal obstruction: total cases, Down's syndrome cases and survival, 1955 – 1985 inclusive

	1955 – 64	*1965 – 74*	*1975 – 85*
Total cases	10 (3)	13 (7)	24 (15)
Down's syndrome	2 (0)	3 (1)	12 (5)

Figures in brackets denote number surviving.

11-year consecutive periods. It will be seen that 12 of the 17 cases of Down's syndrome appeared in the most recent period, presumably therefore giving them the maximum chance of survival. The gradual increase in the number of cases presenting at the Royal Belfast Hospital for Sick Children is probably due to a change in the referral pattern rather than to any increase in the population incidence of duodenal obstruction.

TABLE II

Surgical profile

Site of obstruction	*Pre-ampullary*	*Post-ampullary*	*Annular pancreas*	*Incomplete obstruction*	*No information*
Total	9	30	10	9	8
Alive	8	14	4	9	7

Operation	*Duodeno-duodeno-stomy*	*Duodeno-jejuno-stomy*	*Gastro-jejuno-stomy*	*No surgery*
Total	22	18	4	3
Alive	15	10	0	0

Table II shows the distribution of surgical pathology and the operative management with an indication of survival in each group. Gastro-jejunostomy was performed only prior to 1960. No surgical complication contributed to the death of any patient in the series, and the only early complication was wound infection in three cases.

TABLE III

Duodenal obstruction with or without other congenital abnormalities

Non-Down's group		*Down's group*	
Isolated	With anomalies	Isolated	With anomalies
19 (13)	11 (6)	8 (5)	9 (1)

Figures in brackets indicate number surviving.

The associated anomalies in other systems are shown in Table III. At least one additional abnormality was present in 42.5% of cases. There was only one survivor from nine Down's syndrome babies who had at least one additional anomaly.

DISCUSSION

Our review of duodenal atresia in a captive population over a period of 31 years has shown a gradual improvement in survival, an increase in cases referred, and the early abandonment of gastro-jejunostomy in favour of duodeno-duodenostomy/jejunostomy. The major factor in improved survival of these children lies in the development of neonatal intensive care rather than the development of surgical technique. It was evident that many of the cases in the first 10 years of the study were severely dehydrated on transfer to the specialist centre: aspiration pneumonia and sepsis frequently were complicating factors. These observations have been noted in other larger series.[1] Aspiration pneumonia and sepsis will affect any neonate in whom vomiting is allowed to go unchecked and this brings to light one of the diagnostic pitfalls with duodenal atresia. Preampullary atresia occurs in approximately 30% of cases,[2] and does not produce bile-stained vomitus. Hence babies in this group tend to be managed as feeding problems initially. The gravest situation is when malrotation with midgut volvulus produces duodenal obstruction with the classical 'double bubble' on plain abdominal X-ray. This situation requires urgent laparotomy unlike duodenal obstruction *per se* which demands only elective intervention.[3]

Following surgical correction of duodenal atresia, the proximal duodenum remains dilated and atonic for three to four weeks, so that the anastomosis does not function efficiently. Nasogastric suction must be effective throughout to prevent further vomiting and aspiration, and some workers prefer a gastrostomy in this situation. Feeding is best carried out via a transanastomotic tube.

From a technical standpoint the only complications were three wound infections, which compares favourably with the 15% complications rate reported in many series.[4] Special reference should be made to obstruction associated with annular pancreas which occurs in 25 – 30% cases.[5] No attempt should be made to resect this area since damage to the biliary and pancreatic ducts is inevitable: a standard bypass is therefore indicated.

Similar to other reported series,[1, 2, 4, 5] the Down's syndrome babies comprised 36% of the study and presented with the same distribution of birth weight and gestational age as the rest of the group. The crude mortality was 65% in the Down's sub-group and 47% in the remainder.[5] However, mortality in Down's cases rose to 89% when one other abnormality was present, whilst in the non-Down's group mortality was 66% in the same situation. The 42.5% occurrence of associated abnormalities in the series is similar to that in other studies.[1, 2, 6] This high association of a second and often serious abnormality undoubtedly contributes to the elevated mortality figures.

Management of Down's syndrome babies requiring surgery produces predictable controversy. Attention should therefore be given to their comparative prognosis with non-Down's babies requiring operation. In this area, correction of duodenal atresia and cardiac lesions[7] most commonly cause difficulty, the former because it sets a precedent for treating any one child and the latter because of its magnitude. It is interesting to note that only one correctable renal lesion was

recorded in the Down's group. This was a unilateral hydronephrosis, the other two cases of renal lesions being a single agenetic kidney. Review of the literature shows a 5% association of renal anomalies in duodenal atresia encompassing the full spectrum of urinary tract pathology and therefore tending to be less significant in the management of a Down's baby than duodenal atresia with or without a cardiac lesion.

In this retrospective study, it is evident that, comparing crude and corrected mortality of Down's and non-Down's babies with duodenal obstruction, the Down's group have an overall reduced prognosis. Whilst such an observation should be considered when planning the management of a Down's syndrome baby and when counselling the parents, it does not form a basis for a policy of non-treatment.[8]

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