Congenital Atresia and Stenosis of the Duodenum – A Case Report

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Introduction

CONGENITAL ATRESIA and stenosis are the commonest causes of neonatal obstruction of the small intestines; one or other occurs once in 20,000 births. The sites of the obstruction is as follows (Davis and Poynter):–

Duodenum above the papilla	15 per cent	33 percent
Duodenum below the papilla	18 per cent	
Jejunum	15 ,, ,,	
Ileum	25 ,, ,,	
Colon (usually ascending colon)	10 ,, ,,	
Multiple sites	17 ,, ,,	

We wish to report a case of Mongolism with duodenal atresia and stenosis treated at General Hospital, Johor Bahru recently.

Case Report

A 15 day old male Chinese infant P.N.Y. was refered to the surgical unit General Hospital Johor Bahru from a district hospital as a problem of intestinal obstruction in a newborn on 6th June 1973. It had apparently been a normal pregnancy and delivery for the 39 years old mother of ten children. She had not noticed any marked difference in the appearance of the infant compared to the other siblings when they were babies.

There was a history of vomiting three to four times daily since birth. The vomitus consisted of yellowish liquid mixed with curd. The vomiting was not projectile. Muconeum was passed a few hours after birth and on the third or fourth day of life the muconeum gave place to stools showing the presence of milk curds. The passage of muconeum during the first three days of life does not make negative a diagnosis of intestinal obstruction. For the past four days prior to admission the baby had been constipated. The mother noticed it had been feeding poorly since birth. She had not noticed any abdominal distension.

On physical examination the facies of mongolism was noticed and other confirmatory signs such as short hands with incurved fifth fingers and widely separated first and second toes made a confident diagnosis of Down's syndrome possible. The baby's general condition was satisfactory and there were no signs of dehydration. The cardiovascular and respiratory system were normal. The abdomen was soft with minimal distension of the epigastrium but there were no visible peristalsis observed nor any mass palpated per abdomen. There was diastasis of the recti abdominis muscles. Rectal examination revealed no mass nor any faeces. A plain radiograph supine showed marked distension of the stomach and duodenum and an erect view showed air and fluid levels in the stomach and deodenum. Our preoperative diagnosis was Duodenal Atresia/Stenosis. Differential diagnosis: Annular pancreas.

Preoperatively intravenous infusion and gastric suction were started. Laparotomy was done on 9th June 1973, two days after admission. We found the stomach and the first part of the duodenum dilated beyond a constriction 3 mm. long where the second part of the duodenum began. This

stenosis only admitted the end of a fine probe about 1 mm. in diameter. There were no other sites of obstruction in the jejunum, ileum or the colon. The liver, gall bladder, spleen and pancreas were all normal.

Operation

We decided at operation to perform a DUODENO-DUODENOSTOMY because technically it was possible to do it and we thought it was more physiological than the standard operation of Duodeno-jejunostomy.

An incision was made anteriorly extending from the dilated first part of the duodenum across the stensosis to the collapsed second part of the duodenum. With the anastomosis partially completed with fine 5/9 silk a polythene Ryle's tube was guided through the anastomosis and threaded to the jejumun. The anastomosis was completed in one layer. The patient vomited once on the first post-operative day despite intravenous fluids and gastric suction. The patient became cyanosed and on auscultation there were rhonchi in the lungs. We started postural percussion immediately with tracheo-bronchial suction and this was continued daily for several days. The patient had been on injections Ampicillin. Feeding with clear sterile fluids was commenced on the fourth post-operative day. A gastrograffin swallow was done on the seventh post-operative day to exclude anastomotic leak and also to test the patency of the anastomosis Fig. 1 and Fig. 2. It was reported that the stomach and first part of the duodenum were less dilated and that the dye flowed easily through the anastomosis; there was no leak.

This case was presented at the Combined Monthly Meeting of General Hospital, Johor Bahru on 27th June by Dr. M. Yusof Said.

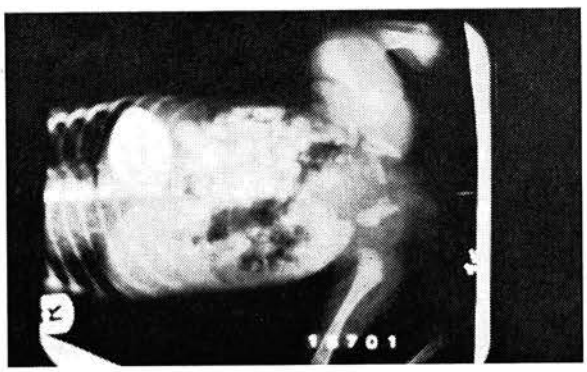

Figure I
Gastrograffin Swallow
Taken on the 7th Post-Operative day, showing patency of Duodeno-Duodenostomy.

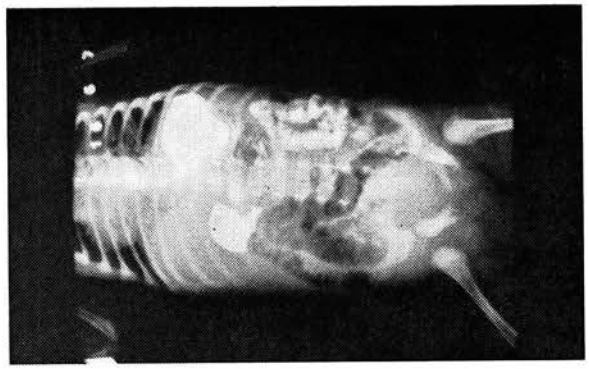

Figure II
Gastrograffin Swallow
Taken on the 7th Post-Operative day.
The first and second parts of duodenum are still dilated.

At the last follow-up in October 1973 the baby was thriving Fig. 3.

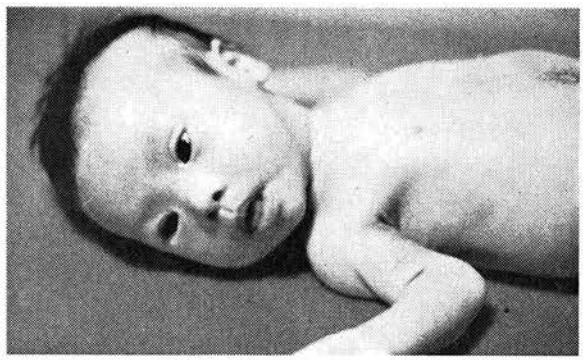

Figure III
Mongol with
Duodenal Atresia/Stenosis Thriving.
3 Months after Operation.

Discussion

Luow (1952) in a study of neonatal obstruction occuring over the period 1926 to 1951 at the Hospital for Sick Children, Great Ormond Street, found 31 reports of cases involving the duodenum. In about 20% of the cases the obstruction was placed proximal to the ampulla of Vater, while in 75% of the cases it was placed distal but close to the ampula of Vater. 5% of cases showed an area of atresia in the third of fourth part of the duodenum. In all cases the duodenum proximal to the obstruction was grossly distended and the walls thinned out to such an extent that rupture could take place.

In the rare condition of duodenal stenosis there is usually a diaphragm present with a small

central perforation, with the result that the intestinal obstruction is not complete. The perforation is usually so small that the symptoms and signs are those of atresia, but if food can pass through, the condition may be difficult to detect. Such an example is presented in this case report.

Symptoms and Signs

As we are dealing with a congenital malformation associated with obstruction, persistent vomiting takes place within the first 24 hours of taking feeds. Vomiting becomes frequent and leads to severe denyhdration and alkalosis and death will ensue within one week unless surgical relief is given. After a feed gastric peristalsis will be seen and the vomit will become projectile in character. Bile will be present in the stomach content in 80% of cases, but where the obstruction lies proximal to or at the level of the ampulla of Vater bile is not present in the vomit, so that delay in diagnosis may occur as the clinical picture is one of mucous gastritis. Normal meconium stools may be passed in the first 48 hours but soon constipation will develop as no food is passing through the duodenum. Farber (1933) showed that squamous epithelial cells derived from the skin of the foetus from the third month of foetal life onwards were ingested in the amniotic fluid and were present in the meconium stools. Failure to demonstrate these cells shows that the intestinal obstruction is complete and that the lesion has been present before the third month.

In atresia a plain radio-graph will show marked distension of the stomach and the duodenum proximal to the lesion, but no air will be seen in the small intestine. The duodenum often is so dilated that radiography suggest that there are two stomachs present. In the rare cases of stenosis; where the size of the lumen is compatible with life, the onset of symptoms of vomiting and constipation is delayed. There is no justification of giving a barium meal; the danger of aspirating vomitus is too great. When it is absolutely necessary to give opaque material (partial obstruction) a small amount of gastrografin given through the gastric tube is useful for demonstrating the location of the obstruction, but the opaque material should be aspirated directly after the films have been exposed.

Differential Diagnosis

Suprapapillary duodenal atresia is distinguished from oesophageal atresia by the fact that there is no dribblling of saliva and there are no attacks of cyanosis after feeding. The absence of a palpable lump serves to differentiate duodenal obstruction from infantile pyloric stenosis and the latter occurs later.

The Surgeon must remind himself that duodenal obstruction in infancy can also be caused by volvulus of the midgut, congenital bands and an annular pancreas.

Lastly, attention is directed to the high incidence of mongolism amongst sufferes from duodenal atresia and stenosis. White, Carter and Luow (1952) found a high incidence of mongolism, 10 out of a consecutive series of 32 cases being affected.

Treatment of Duodenal Atresia

Intravenous infusion is given to restore the electrolyte imbalance and the state of dehydration set up by severe vomiting. Gastric suction is set up prior to operation to prevent the aspiration into lungs of fluid secretions that pour into the distended duodenum and stomach.

Anaesthesia

General anaesthesia with the use of nitrous oxide, oxygen and other, is the anaesthetic of choice. The metabolic rate of the infant is high and respiration is often depressed due to a state of alkalosis, so a high percentage of oxygen has to be given. General anaesthesia, administered by an anaesthetist experienced in anaesthetizing infants, by providing relaxation of the abdominal wall, simplifies the surgeon's task. Endotracheal anaesthesia provides a free airway in the presence of an unstable respiratory centre, and prevents inhalation of gastric contents.

Probably the safest, by no means the most satisfactory is local infiltration with oral sedation. When local anaesthesia is employed some form of restraint is necessary; the best method is to bandage the wool-covered extremities firmly to a padded crucific.

Operation

Retrocolic duodenojejunostomy is the operation of choice. The mortality has been reduced by the following small addition to the operation. After completing the anastomosis, a stab incision is made through the wall of the pyloric anturm. Through this incision a plastic catheter is passed, and its tip is guided through the anastomosis. The tube is anchored to the stomach wall by a catgut stitch, and a valvular opening is constructed. The tube is brought to the surface either through a stab incision or through the upper part of the laparotomy incision, whichever gives the more direct passage. This additional step allows rest to the duodenum until its muscle tone has recovered, when molk feeding can be commenced through the tube. Gastrojejunostomy should not be used for this condition. During the anastomosis the intestinal content is removed by suction and clamps are not applied

as they may devitalise the thinned out duodenal wall. A single layer of interrupted fine silk sutures will complete the anastomosis.

Ehrenpreiss and Sanblom (1949) claim to have reduced their operative mortality from 75% to 25% by intubation of the anastomotic opening.

Post-operative management based on the knowledge that the dilated duodenum will not regain tone or true peristalsis for several days will lead to further reduction in operative mortality.

Acknowledgements

We wish to thank the Director General of Medical and Health Services of Malaysia, Tan Sri Datuk (Dr.) Abdul Majid Ismail for permission to publish this case report and Cik Siti Mariam and Cik Salmah for typing it.

References

1. Ehrenpreis, T. and Sandblom, P. Acto Paediat, Uppsala 38, 109 1949.
2. Farber, S., J. Amer, Med. Ass., 100, 1753, 1933.
3 Gross, R. E., The Surgery of Infancy and childhood, Saunders, London and Philadelphia, 1953.
4. Luow, J. H. and Barnard, C. N. Congenital Intestinal Atresia: Observations on its Origion. Lancet 2: 1065, 1955.
5. Luow J. H. Congenital Intestinal Atresia and Stenosis in the newborn. Observations on its pathogenesis and treatment. Ann. Roy. Coll. Surg. 25: 269, 1959.
6. Masen Brown J. J. Surgery of Childhood Edward Arnold (Publishers) Ltd. 1962.