Summary:
We present here a 7 years' old boy presented with recurrent abdominal pain and vomiting causing severe growth failure since 6 months of age. He had some dysmorphism including fish like mouth, upturned nostrils, hypertelorism, low set posteriorly rotated ears, absence of left kidney and closing ventricular septal defect (VSD). Barium study revealed extrinsic obstruction in duodenal 3rd part with high up caecum. Laparotomy showed Ladd's band. Ladd's procedure was done upon this child, which led to resolution of his clinical abdominal problems.  

Case Summary

Joy, a boy of 7, from Dhaka was admitted in Bangladesh Medical College Hospital on 02.07.2005 with recurrent upper abdominal pain and vomiting with severe wasting since 6 months of age. His grandmother said that he was growing almost uneventfully up to 6 months of age. Each attack of abdominal pain and vomiting usually lasted for 4-5 days. The problem recurred in an almost similar pattern once in every 2-3 weeks. The pain was localized, moderate to severe occurring around the umbilicus, comes and goes, and aggravated by food intake. Occasionally he feels a dull aching pain in both loins. During each attack he vomits several times mostly after meals. Vomiting is not projectile, and contained only food particles, no blood, and no bile. He gets severely constipated during each episode. Joy did not have any urinary complaints. Parents were concerned, as their child was not growing rather getting stunted and wasted. He was admitted at least 20 times in different hospitals till to date. Once he was admitted in a hospital where he was investigated and suspected to be a syndromic child with congenital absence of left kidney, VSD and gastro-esophageal reflux disease (GERD). Joy was born full term normally at home. Mother had regular antenatal care and the pregnancy was uneventful. He cried just after birth and sucked breasts strongly. Feeding was otherwise normal. Joy was immunised. His growth was normal for the first 6 months. Developmental milestones were satisfactory.

Joy is the only child of his nonconsanguineous parents. Other family members are reasonably well. Physically, Joy was looking grossly emaciated and stunted. His weight was 10 kg (35% of reference value of CDC; far...
below the 3rd centile), height 95 cm (71.2% of reference value of CDC, far below the 3rd centile), Occipitofrontal Circumference (OFC) 44 cm (reference value for 6 and half months according to CDC); and Mid Arm Circumference 14 cm. His psychomotor, visual, hearing and speech development was absolutely normal. Joy was looking dysmorphic with fish like mouth, upturned nostrils, hypertelorism, low set posteriorly rotated ears, looking pale but no jaundice, oedema or enlarged lymph nodes. Pulse was 68/min, temperature 98°F and breathing 16. He was alert and cooperative. Heart showed normal S1 and S2 with no murmur. Chest was normal. There was no abdominal distension, tenderness, nor any organomegaly. CNS examination revealed normal findings. Laboratory data following admission shows normal peripheral blood count and film study; normal liver functions, and urinalysis. Electrolytes and renal functions were normal. USG revealed no abdominal organomegaly with absence of left kidney. DTPA shows absence of left kidney with normally functioning right kidney. IVU shows non-visible left kidney with normal right kidney. Color Doppler echocardiography done earlier (2004) showed two small perimembranous VSD (almost closing), mild Tricuspid Regurgitation with normal pulmonary arterial pressure. Barium meal and follow-through showed eccentric narrowing of 3rd part of duodenum from outside with thicker mucosal pattern of upper jejunum and high up caecum.

Fig-2: Barium meal of upper GIT shows narrowing of the 3rd part of duodenum.

Fig-3: Ba-meal and follow-through of upper GIT shows high up caecum.

Fig-4: Shows peroperative view of Ladd's band.

Fig-5: Shows Ladd's band
Laparotomy was done and revealed normal stomach but dilated first and second duodenal parts. Caecum was high up, almost under the gallbladder. There were lot of bands between caecum and right paravertebral gutter. The band was carefully divided. Kocherization of duodenum done. The caecum was transferred to left iliac fossa and appendicectomy was carried out. Postoperative recovery was uneventful. He went back home with advice to take small frequent meals. Zinc, iron, and multivitamins were supplemented. Joy was advised to report after 2 weeks. Joy reported after 1 month with no abdominal complaints and gained a weight of 2 kilogramms.

Discussion
Extrinsic obstruction of the duodenum may be caused by congenital peritoneal band, intestinal malrotation, annular pancreas, preduodenal portal vein and duodenal duplication. Peritoneal band is always associated with intestinal malrotation, which means an incomplete rotation of intestine during fetal development. There are 4 types of congenital peritoneal band. In type 1, caecum that lies abnormally in right upper quadrant of the abdomen, has a band, which extends across the 2nd and 3rd parts of the duodenum to the paravertebral gutter, this band is called Ladd’s Band. Duodenal obstruction may result from either compression by Ladd’s Band and/or from midgut volvulus. The type 2 band extends from the hepatic flexure of the colon across the 2nd part of duodenum to the right paravertebral gutter, causing duodenal compression at that site. The type 3 band is the hypertrophied hepatoduodenal ligament, which obstructs the duodenum at the junction of its 1st and 2nd parts. The type 4 band is a dense fibrous band which binds the distal portion of the 3rd part of the duodenum to the paravertebral fascia, causing extrinsic obstruction and is always associated with an incompletely rotated duodenum.

It seems that this case falls in the type 1 category.

Clinical diagnosis of congenital band with malrotation in older child is not always easy because of its non-specific presentation. Commonest complaints are intermittent colicky abdominal pain (100%), recurrent vomiting (89%), haematemesis and constipation (55%). Repeated episodes of bloody stool, diarrhoea and failure to thrive (44%). In all cases of persistent vomiting specially biliary a contrast Ba-meal and
follow-through study should be done which is diagnostic if correctly interpreted. 6,7 It should be remembered that congenital peritoneal band is always associated with malrotation and this can cause volvulus of the midgut. In one series 31 out of 219 children had volvulus, 7 needing resection with one death.8, 9 Therefore, once band is diagnosed Ladd’s procedure should be routinely done. Nowadays, it can be done by laparoscopy having the advantage of short convalescence and low morbidity.10, 12 The association between gastro-esophageal reflux (GER) and intestinal malrotation has well been described. Delayed or impaired gastric emptying is thought to be a contributing factor in the development of GER.11 This child was having persistent periodic vomiting and got different brands of anti-reflux drugs with little benefit. This should have had raised the query to clinician’s mind about the possibility of malrotation. Recurrent pancreatitis is also described in malrotation by some authors and this resolved spontaneously after Ladd’s Procedure.12 This child with some dysmorphic features having absent left kidney, closing VSD, recurrent abdominal pain and vomiting eventually developed severe growth failure and protein energy malnutrition. Single malformation in one system should warn the clinician to have a look for anomalies into other systems. A simple investigation like barium-meal and follow-through would have solved this child’s problems much earlier. Any child with long standing periodic vomiting with recurrent abdominal pain causing growth failure an upper GIT barium study should be a routine procedure.

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References