Introduction
There is very high incidence (36% or one third) of congenital duodenal obstruction in Down’s syndrome [1-2]. Congenital duodenal obstructions present with bilious vomiting and are relatively uncommon to be proximal to the ampulla of Vater about 30%, the most common site being peri-ampullary [2]. Pre-ampullary lesions with non-bilious vomiting simulating gastric outlet obstruction has been reported recently [1]. We describe a case in which despite high index of suspicion, the diagnosis of distal duodenal stenosis close to duodeno-jejunal flexure eluded prenatal and postnatal detection posing diagnostic and therapeutic challenges.

Case Report
A first baby boy was born by normal vaginal delivery at 30+5/40 weeks with birth weight 1750 grams having trisomy 21 without polyhydramnios. He had mild respiratory distress syndrome requiring nasal continuous positive airway pressure for six weeks in special care baby unit. There was systolic heart murmur in neonatal period and echocardiography showed small ventricular septal defect which closed spontaneously. He passed meconium in first 48 hours but then required daily suppositories to open bowels. He had intermittent non-bilious vomiting up to 20 times a day, constipation with bowels opening every 4 to 5 days and occasional bilious aspirates. On examination, the abdomen was soft, non-distended and non-tender. Abdominal film showed normal bowel gas pattern in the immediate postnatal period while on continuous positive airway pressure (CPAP) (Fig. 1A).

With the help of suppositories, he had explosive bowel movements and Hirschsprung’s disease was suspected. He therefore underwent rectal suction biopsy which however showed plenty of normal ganglion cells. He was thus commenced on laxatives. Despite this he continued to have same symptoms and subsequently underwent abdominal films showing stomach and duodenum moderately distended but distal bowel gas visible. He previously had normal bowel gas distribution while he was on CPAP, for these reasons an upper gastrointestinal study was not suspected (Fig. 1B-1D).

Abstract
We present a case of distal duodenal stenosis without a wind sock deformity in a Down’s syndrome presenting with non-bilious vomiting, constipation and bilious aspirates simulating Hirschsprung’s disease. Despite high index of suspicion and actively looking for associated congenital duodenal obstruction it eluded detection. The plain film and rectal biopsy in neonatal period were normal. It was the upper gastrointestinal study which helped in diagnosis. At exploration, distal duodenal stenosis without wind sock deformity was identified. Duodeno-jejunostomy was curative.

Keywords: duodenal stenosis, Down’s syndrome, duodeno-jejunostomy
He continued to have constipation, failure to thrive with centile dropping from 9th to 2nd centile with frequent vomiting 6 to 12 per day despite full antireflux medications. It was now noted that on occasions, vomits were bile stained. He therefore underwent upper gastrointestinal contrast study. This showed dilatation of the stomach and duodenum with delayed transit of contrast through a significantly narrowed third part of duodenum with a distinct change of caliber and without evidence of malrotation. The appearances did not appear typical of a duodenal web and duodenal stenosis or a ‘wind-sock’ was more thought to be more likely (Fig. 2).

He underwent exploratory laparotomy at 3 months corrected age. At exploration, there was grossly dilated stomach and duodenum to the third part and there was calibre change at the junction of 3rd part with the 4th part of duodenum but no evidence of a web or external band. The duodeno-jejunal flexure was normally positioned. There were multiple lacteals throughout the entire small bowel mesentery. The options of Strong’s manoeuvre converting normal rotation into malrotation, duodenoduodenostomy (technically impossible as the stricture was distal duodenal), and duodeno-jejunoscopy were considered. On balance, the last alternative was selected. A transverse incision was made on the dilated duodenum and the absence of a diaphragm or web was confirmed. A feeding tube size 8 could not be passed into the 4th part of duodenum. A longitudinal incision was made on the proximal jejunum just beyond the flexure and tension free duodeno-jejunoscopy was carried out uneventfully. No trans anastomotic tube was employed.

His initial post-operative period was uneventful and then he had nasogastric tube aspirates ranging from 400 to 750 ml in the following few days. A decision to start total parental nutrition was made and he underwent Broviac line insertion under general anaesthesia. The aspirates started falling in the second postoperative week. He was regaining bowel movements and feedings were started on 12th post-operative day which he tolerated slowly and steadily and was discharged home on 16th post-operative day. At follow up at 3 months he was thriving well and was on anti-
reflux medications. At 1 year, he is progressing well and weaning from his antireflux medications.

Discussion

Complete duodenal obstruction is often associated with polyhydramnios. This important clue could be absent in partial duodenal obstruction with stenosis. There is a very high incidence of duodenal anomalies in Down’s syndrome and although prenatal team was actively looking for it, the partial obstruction could not be detected antenatally. In duodenal atresia, passage of meconium at birth is exceptional and invariably a very good clinical indicator of the underlying complete obstruction [2]. Our patient passed meconium in the first 48 hours but then developed continued constipation. When the initial plain film was taken, the baby was on nasal CPAP, the gas ingested was significant and so was the distribution along the entire gut. This misled us to the possibility of Hirschsprung’s disease requiring rectal suction biopsy. It also misled the radiology team as each time reference was made to previous normal bowel gas pattern thus delaying early detection.

Congenital duodenal obstructions presents with bilious vomiting and are relatively uncommon in the distal duodenum, the most common site being just at the ampulla. Presentation with hematemesis has been reported [4]. Failure to thrive during weaning period may unmask the underlying partial obstruction as happened in our case [5]. The pitfalls in the diagnosis of partial duodenal obstructions have been described by previous authors [6-10]. Partial duodenal obstruction is known to present later on in life [11]. Plain radiographs that demonstrate the double-bubble appearance with no distal gas is characteristic of complete duodenal obstruction but not of stenosis or partial obstruction. The important lesson from our case was that even in the absence of bilious vomiting, an upper gastrointestinal contrast study is the gold standard to detect partial obstructions and that there needs to be a low threshold in performing this study. Upper gastrointestinal contrast studies will delineate the nature and site of the obstruction. Presence of a dimple on fluoroscopy suggests possibility of a duodenal web [12].

The distal duodenal stenosis was not only a diagnostic challenge with all the odds as mentioned above but has several therapeutic challenges and difficult options. Theoretically duodenal stenosis can best be treated with duodenoplasty with a longitudinal incision along
Isoperistaltic duodeno-jejunostomy entails a larger chylous ascites and postoperative adhesions. Taking and is associated with prolonged ileus, normal rotation into non rotation is a major undertaking and is associated with prolonged ileus, chylous ascites and postoperative adhesions. Isoperistaltic duodeno-jejunostomy entails a larger loop of 4th part of duodenum and proximal jejunum predisposing to blind loop syndrome and malabsorption with bacterial overgrowth. Under the circumstances we selected antiperistaltic loop directly anastomosis which led to increased aspirates temporarily in the postoperative period. The passage of trans anastomotic tube above the anastomosis may have helped in starting the nasojejunal feeds early and may have helped avoiding insertion of the central line as shown by recent studies in reducing the need for the central line and total parenteral nutrition [15-18]. In conclusion, distal duodenal stenosis in association with Down syndrome could be a diagnostic and therapeutic challenge despite high index of suspicion. We recommend actively look for partial obstruction and if the baby is on CPAP, the interpretation of the plain film, particularly the gas distribution should be with caution. The upper gastrointestinal contrast study remains the gold standard for early diagnosis and threshold should be low. Once diagnosed, all available surgical options should be considered and the best one implemented.

REFERENCES