International Surgery Journal
Chandrasekaran S et al. Int Surg J. 2017 Aug;4(8):2605-2611
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: http://dx.doi.org/10.18203/2349-2902.isj20173396

Original Research Article

Clinical profile and predictors of outcome in congenital duodenal obstruction

Sankkarabarathi Chandrasekaran[1], Anirudhan Asokaraju[2]*

[1]Department of Paediatric Surgery, Madras Medical College, Chennai, Tamil Nadu, India
[2]Department of Paediatric Surgery, Stanley Medical College, Chennai, Tamil Nadu, India

Received: 05 June 2017
Accepted: 19 June 2017

***Correspondence:**
Dr. Anirudhan Asokaraju,
E-mail: drcsbarathi@yahoo.co.in

ABSTRACT

Background: Congenital duodenal obstruction is a broad term that refers to a variety of disorders like duodenal atresia, duodenal stenosis, annular pancreas, duodenal membrane and preduodenal portal vein. The aim of this study was to analyse in detail the entire clinical profile of duodenal obstruction and to study the short-term outcome.
Methods: A prospective descriptive study was conducted in the Department of Paediatric Surgery, Institute of child Health and Hospital for Children, Egmore, Chennai, among all babies with suspected duodenal obstruction based on clinical symptoms and subsequently proven by surgery from September 2013 to January 2015. Fifty babies were selected for this study based on the eligibity criteria.
Results: Upper gastrointestinal contrast radiography is the gold standard investigation in diagnosing duodenal membrane with a hole. The type of surgery such as the duodenoplasty, duodenoduodenostomy or duodenojejunal anastomosis did not affect the post-op outcome.
Conclusions: Long term follow up is warranted to know the actual incidence of requirement for redo procedure for feed intolerance. The presence of coexisting Down's syndrome and congenital cardiac lesions increases the mortality rate in these children.

Keywords: Congenital duodenal obstruction, Duodenal atresia, Down's syndrome

INTRODUCTION

Congenital duodenal obstruction is a broad term that refers to a variety of disorders like duodenal atresia, duodenal stenosis, annular pancreas, duodenal membrane and preduodenal portal vein.[1] The Kimura's procedure described in 1977 is the standard procedure for re-establishing GI continuity.[2] The diagnosis is straight forward in a newborn presenting with bilious vomiting and a typical double bubble appearance in a plain skiagram. Prenatal scans account for the antenatal diagnosis of some cases. The coexistence of trisomy 21 and other cardiac anomalies such as septal defects add to the morbidity.[3] The time of presentation, different surgical techniques and their effects on outcome of these neonates has also been discussed in recent literature.

Thus, the authors planned and designed this study with the following aims and objectives. The primary aim was to analyse the entire clinical profile including epidemiology, clinical presentation, operative findings, post-operative complications and short-term outcome of congenital duodenal obstruction in neonates.

The secondary objectives were to analyse the value of imaging in diagnosing diverse types of duodenal obstruction and also to study the incidence of associated anomalies with them. The presence of trisomy 21 or

cardiac anomalies and their effects on outcome was also studied.

METHODS

A prospective descriptive study was planned to conduct in the Department of Paediatric Surgery, Institute of child health and hospital for Children, Egmore, Chennai, among all babies with suspected duodenal obstruction based on clinical symptoms and subsequently proven by surgery from September 2013 to January 2015. Fifty babies were selected for this study based on the eligibity criteria.

Inclusion criteria

Neonates with or without antenatal scans and presenting with bilious vomiting at birth and laparotomy proven duodenal obstruction within this study period.

Exclusion criteria

Patients with malrotation and other forms of GI atresia were excluded.

Babies presenting with bilious vomiting at or soon after birth were admitted and prospectively evaluated for the cause of vomiting. X-ray abdomen, USG abdomen in all patients and barium meal study in some of the doubtful cases were done. Clinical profile was analyzed by taking detailed history regarding consanguinity, antenatal scans, polyhydramnios, premature birth, comorbidities like Down's syndrome and family history. Detailed clinical examination was performed and operative procedure depending on the cause of duodenal obstruction like atresia, membrane with whole or annular pancreas was done. The role of the time of diagnosis, patient factors, time of intervention, operative techniques on the post-operative outcome was studied.

Investigation protocol

Babies with antenatal scan suggestive of polyhydramnios and dilated stomach presenting with bilious vomiting are initially resuscitated with IV fluids and nasogastric tube inserted. A plain X-ray abdomen was taken and if it showed "double bubble" sign, the neonates were taken for emergency laparotomy and proceed.

If the newborn was clinically stable with no antenatal scans and the plain skiagram was not contributory, they were subjected to an USG abdomen with colour Doppler. The radiologist specifically looks for the dilated stomach and proximal duodenum and collapsed loops beyond that. An altered SMA/SMV axis is often noted suggestive of a coexisting malrotation with duodenal obstruction. The USG scan also picks up conditions like annular pancreas. Rarely, a pre-duodenal portal vein may be identified in a doppler study. If the clinical symptoms were consistent with duodenal obstruction and ultrasound doppler abdomen features are suggestive, the patient was again taken up for exploration. If either the clinical scenario or Ultrasound was equivocal then the patient underwent an upper gastro intestinal contrast study.

Barium meal follow through (BMFT) is the contrast study of choice used in our institute for doubtful cases. Early AP/PA/RAO films show the size of the stomach and duodenum, position of DJ flexure in relation to the vertebral body and the gastric outlet, and the side of the early jejunal loops. Late films show the position of caecum. The BMFT mainly diagnoses other causes of duodenal obstruction such as the ampullary membrane with a hole.[4] There is dilated stomach and duodenum with a narrowing at the second part and trickling of contrast down the lane.

The "wind sock" sign is a special condition where the duodenal membrane at the ampulla bulges down if the third or fourth portion of the duodenum and shows apparent obstruction distally. This sign is also suggestive of a duodenal membrane with a hole.

Surgical management

All babies underwent the standard Laparotomy and Kimura's diamond shaped duodeno-duodenostomy for correction of duodenal obstruction. The newborn with duodenal membrane underwent duodenotomy, membrane excision and closure. A tapering duodenoplasty was done occasionally in a hugely dilated proximal duodenum. Rarely, asymptomatic patients with positive radiologic findings, a diagnostic laparoscopy was contemplated to decide upon the need for surgery.

RESULTS

The study comprised of fifty babies who were admitted and operated for congenital duodenal obstruction from September 2013 to December 2015 who satisfied the inclusion and exclusion criteria.

Table 1 presents demographic and baseline characteristics of the babies. Congenital duodenal obstruction commonly presents in the new born period, with 84% (42) in this study presented on the first day of birth followed by 12% (6) presented within the first week and 4% (2) within the first year of life. In this study, congenital duodenal obstruction appears to be slightly more frequent in female children (28 patients) than in males (22 patients) in the ratio of 1.27:1. Among the 20 term babies, 13 were appropriate for gestational age with a birth weight of more than 2.5 kg and out of the 30 preterm babies 22 were more than 1.5 kg. Consanguineous marriage was found in only 22 (44%).

Antenatal scan finding of polyhydramnios and double bubble appearance are suggestive of duodenal obstruction. In this study, only 12 patients had an antenatal diagnosis of duodenal obstruction. One patient

had undergone a fetal MRI which was suggestive of duodenal atresia.

Table 1: Demographic and baseline characteristics.

Age	Number (%)
<1 day	42 (84)
1 week	06 (12)
<1 year	02 (4)
Gender	
Male	22 (44)
Female	28 (56)
Birth weight of term neonates (n=20)	
>2.5 Kg	13 (65)
2-2.5 Kg	6 (30)
<2 Kg	1 (5)
Birth weight of preterm neonates (n=30)	
>1.5 Kg	22 (73.3)
<1.5 Kg	8 (26.6)
Consanguinity	
NON-CM	28 (56)
II Deg	12 (24)
III Deg	10 (20)

The symptoms of the patients were documented and according to their commonly shown symptoms they were distributed. The reports are given in Table 2. Bilious vomiting at birth and inability to feed was the most common symptom observed in 41 patients (82%). 5 patients presented primarily with symptoms of delayed vomiting and subsequently diagnosed to have duodenal membrane after barium meal study and confirmed during surgery. Extreme low birth weight and prematurity can result in an asymptomatic baby even with a significant duodenal obstruction and an incidental finding of duodenal atresia. Among the 48 neonates 42 of them presented within 1 day of onset of symptoms and 3 patients presented within 3 days. Two babies presented beyond the newborn period. One was a case of annular pancreas and was found during evaluation for nonbilious vomiting. The other child was a case of duodenal membrane with a hole and presented a 3rd month of age with failure to thrive.

Table 2: Patients distributions according to symptoms (N=50).

Symptoms	Number (%)
Bilious vomiting	41 (82)
Asymptomatic	1 (2)
Failure to thrive	1 (2)
Delayed vomiting	5 (10)
Abdominal distension	2 (4)
Duration (N=47)	
1 Day	42
3 Days	3
4 Days - 1 Week	3

X-ray abdomen was suggestive of duodenal obstruction if a dilated stomach and dilated duodenum with relative paucity of air or absence of air in small and large bowel was found. Double bubble sign, although suggestive of duodenal atresia, when associated with bilious vomiting a differential diagnosis of malrotation was also considered. The radiological study reports are shown in Table 3. Out of the 50 patients in whom the primary diagnosis was congenital duodenal obstruction, x-ray abdomen was suggestive in 35 patients (70%). Of note is the appearance of gas in the small bowel, which was absent even in the doubtful cases. The presence of dilated stomach should also alert the surgeon regarding the possibility of duodenal obstruction in the neonate.

Table 3: Radiological studies of patients (N=50).

X Ray	Number (%)
Suspicion	02 (4)
Normal	06 (12)
Doubtful sign	35 (70)
Dilated stomach only	07 (14)
Small bowel loops	00
USG abdomen done	
Duodenal obstruction was picked up in almost 90% of the cases	45 (90)
Malrotation	6
Genitourinary	2
Duodenal Stenosis	1
With anorectal malformation	1
Preduodenal portal vein	1

Among the 35 patients with suggestive X-ray, duodenal obstruction was confirmed at the time of surgery in all the cases and was invariably due to duodenal atresia except for 2 cases. Double bubble sign was present in 35 patients. All 35 of them were taken for laparotomy after an USG abdomen. Thirty-two had intrinsic duodenal obstruction and underwent Laparotomy and Kimura's duodeno-duodenostomy. Two patients had malrotation with preduodenal portal vein and one patient had duodenal stenosis. All fifty patients had an ultrasound abdomen even though some had an antenatal diagnosis and plain skiagram finding of double bubble appearance. This was done to identify coexisting anomalies like malrotation in some babies and other genitourinary anomalies. SMA/SMV axis was altered in 6 patients (12%) making ultrasound a sensitive tool in diagnosing malrotation as a coexisting anomaly. One of the 6 patients also underwent an upper gastrointestinal contrast study to confirm malrotation whereas the other 5 patients were operated based on ultrasound findings alone.

Among the 50 patients who underwent USG abdomen, 45 had positive ultrasound findings (90%) and only five patients (10%) required further investigations for diagnosis. As stated earlier, one patient initially diagnosed as duodenal atresia was found to be a case of duodenal stenosis and another turned out to be a

preduodenal portal vein. Thus, USG abdomen is a sensitive tool for diagnosing duodenal obstruction in neonates.

Genitourinary anomaly was found in 2/50 patients accounting for roughly 4% of cases. Associated anorectal malformation was present in one patient roughly 2% of cases. The sensitivity of USG abdomen in diagnosing coexisting anomalies is represented in the above shown bar diagram. Rarely USG abdomen also picked up one case of duodenal stenosis preoperatively in this study.

Barium meal follow through (BMFT) is the investigation of choice for patients with suspected duodenal membrane with a hole. The presence of a dilated stomach and duodenum with a tapering at the level of second part of the duodenum is a characteristic appearance in an upper GI contrast study. Beyond the level of obstruction, there will absence of barium or there will only be a trickling of barium in to the distal small bowel.

BMFT was done in a total of 07 patients (14%). Barium enema was done in one baby with suspected associated hirschsprungs disease. There was no transition zone demonstrated in that patient. CT abdomen was not required for diagnosis in any of the neonates. The results of contrast radiograph are summarized Figure 1. Contrast study is 100% sensitive in detecting duodenal membrane with hole. All the 5 patients in our study were diagnosed preoperatively by a barium meal follow through study. The BMFT study also diagnosed 3 patients with coexisting malrotation preoperatively. It thus accounts roughly for 50% of the cases of coexisting midgut malrotation.

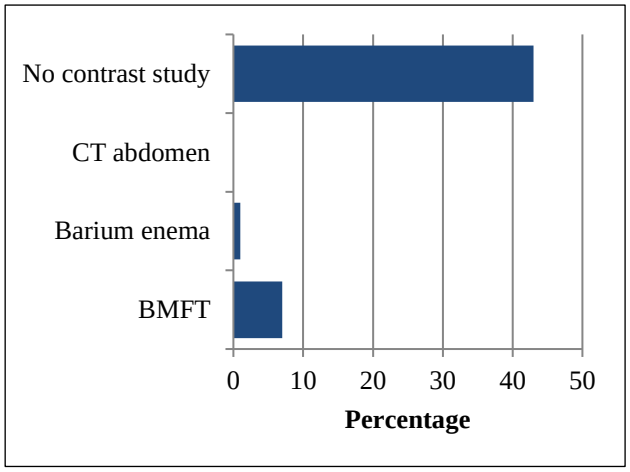

Figure 1: Contrast radiograph.

Kimura's diamond shaped duodeno-duodenostomy procedure was done in open method in all 45 patients. In patients with duodenal membrane, duodenotomy, membrane excision and duodenotomy closure was done. The intra-operative findings confirmed the pre-operative diagnosis in most of the patients.

Two patients underwent surgery in the immediate post-operative period. One was done for persistent vomiting after feeds and diagnosed as persistent feed intolerance. Another patient was operated for leak from the anastomotic site and a re-look laparotomy was done and the baby underwent reanastamosis and survived.

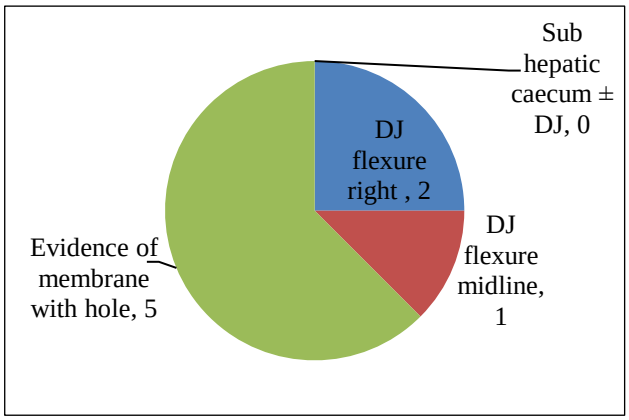

Figure 2: BMFT findings (N=7).

Mortality rate was 12% as 6 patients died during treatment. The results are shown in Table 4. The primary diagnosis in them included duodenal atresia and Down's syndrome in 4 patients, Membrane with hole only in 1 patient and presented late with severe dehydration and shock. One new born had preduodenal portal vein with large ventricular septal defect. The cause of death was sepsis and shock in 1 patient, congenital heart disease in 5 patients of which four patients had associated Down's syndrome.

Table 4: Operative findings (N=50).

	Number
Anomaly	
Duodenal atresia	35
Annular pancreas	06
Duodenal membrane with hole	05
Duodenal stenosis	02
Preduodenal portal vein	02
Death (N=6)	
Duodenal atresia with Down's syndrome	4
Duodenal membrane with a hole	1
Preduodenal portal vein	1
Readmission (N=9)	
Feed intolerance	5
Medical causes	2
MCDK evaluation	1
ARM surgery	1

Excluding the 6 patients who died, the average duration of stay for the patients who survived was 10 days. The duration of stay ranged from 6 to 44 days. A total of 9 patients (18%) were readmitted for various causes. 5 of them had symptoms of feed intolerance (10%), of which

one got operated. This patient underwent a duodenoplasty of the large ecstatic duodenum. This baby improved well subsequently. 1 patient was admitted for evaluation of urological anomaly (MCDK), 1 patient for completion of anorectal malformation surgery. 2 other patients were readmitted for medical causes.

DISCUSSION

Congenital duodenal obstruction was extensively evaluated in 1929, when Kaldor published his experience and identified nearly two hundred and fifty patients with duodenal atresia.[5] However at that time there were only totally nine survivors. This study is the first in our institute aimed at evaluation of all the variables from epidemiology to management of congenital duodenal obstruction.

Congenital duodenal obstruction found in the new born period, with 84% in this study. Similar reports were published previously.[6] The incidence of duodenal atresia roughly varies between 1:6000 to 1:10000 of the live births as per the literature.[7] There is no genetic anomaly or gene defect identified to be associated with duodenal atresia so far. The frequent association with the Down's syndrome and the sibling studies show that there is a likely relationship with a single gene defect that is yet to be identified. Comparable results were reported by Choudhry et al.[8] In the present study we survival rate was 88%. But Choudhry and co researchers found 96% survival rate in 65 neonates with duodenal atresia.[8] However, one out of two of the deaths in our study were due to late presentation in a baby with duodenal membrane and another in a newborn with preduodenal portal vein and multiple congenital anomalies. Hence, a direct comparison with the other studies cannot be made. Only an inference can be taken.

In this study, the cause of death in case of membrane with hole is the very late presentation. The baby had already gone in for severe dehydration and shock prior to resuscitation and followed a spiral downhill course after the surgery. The coexisting cardiac anomalies were the cause of death in babies with coexisting Down's syndrome.

Sarin et al has published a large series of duodenal web in the neonatal surgery journal in the year 2012.[9] In comparison to the present study, they had reported a total of 18 neonates against our statistics of 5 patients. The mean age of presentation of duodenal membrane in our study was roughly 5 days after birth which was in comparison to that the study by Sarin et al.[9]

Some probable causes for delay in clinical presentation include most of the cases were referred from remote villages, the absence of facilities of good antenatal scans and early feed tolerance on the first day has also led to the late referral. Two patients referred late did not have a classical plain X-ray feature of "double bubble" appearance. In such case instilling about 30 to 40 ml air and taking the X-ray film can help in clinching the diagnosis. Lack of parental education and awareness also has led to the delay in presentation of these cases. The delayed presentation is also notorious for increasing the mortality rate.

The Kimura's diamond shaped duodeno-duodenal anastomosis is a wide anastomosis where a transverse incision is made in the proximal dilated duodenum and a longitudinal incision is made in the distal collapsed duodenum. In another technique, the proximal duodenum is partially excised and tapered with a longitudinal suture line to allow early peristalsis. This is proven to avoid the complication persistent feed intolerance following the conventional procedure. This procedure was also done for one baby.

Duodenojejunal anastomosis was previously done before the advent of duodeno-duodenostomy. This is usually avoided as it creates a big blind loop. However, in an extreme preterm infant it can be done. In the present study, it was done in one patient. All babies with duodenal membrane underwent duodenotomy, membrane excision and duodenotomy closure.

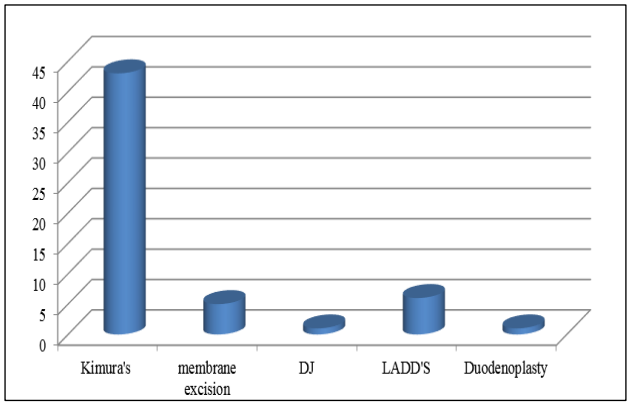

Figure 3: Type of surgery.

Seven of the babies in our study were found to have coexisting malrotation and underwent Ladd's procedure in addition. Post-op feed intolerance is an irritable complication both to the neonate and the operating surgeon. There is no clear role for duodenoplasty in all cases and management protocol is still evolving.[10] Mortality rate was 12% as 6 patients died during treatment. The primary diagnosis in them included duodenal atresia and Down's syndrome in 4 patients, membrane with hole only in 1 patient and presented late with severe dehydration and shock. One new born had preduodenal portal vein with large ventricular septal defect.

The survival rate for these anomalies which was dismal previously has improved in the recent days and has reached up to 95%.[11] In the present study also including only cases of duodenal atresia, the mortality rate is only

8% which is comparable with the international standards. There are some factors such as lack of elective ventilators for some patients, lack of one to one nursing care and complete aseptic precautions which also add to the mortality in these newborns undergoing such major surgeries.

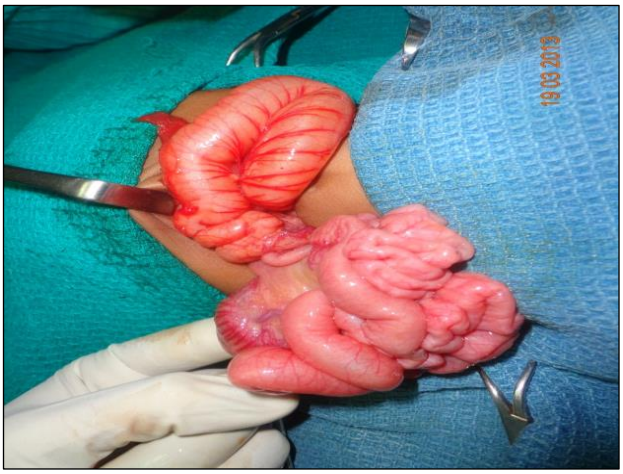

Figure 4: Malrotation with midgut volvulus and viable bowel.

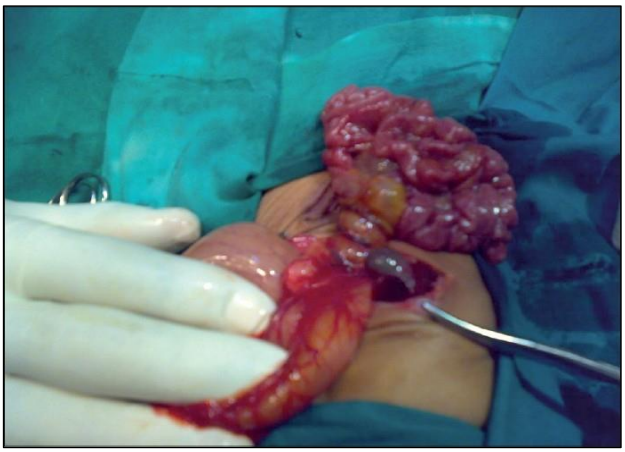

Figure 5: Malrotation with midgut volvulus and gangrene.

Hence, giant strides are needed in the infrastructure, health spending, and commitment of the medical and paramedical staff and improved equipments in the newborn surgical care to take the present challenging work much forward.

CONCLUSION

Duodenal obstruction is a common cause of intestinal obstruction in newborns. Duodenal atresia presents more commonly in neonates, especially in the first day of life as an acute emergency. Bilious vomiting is the most common presenting symptom. A plain x-ray abdomen with dilated stomach and duodenum with paucity of air shadow distally in symptomatic patients is usually associated with duodenal obstruction. Malrotation with volvulus had to be ruled out.

Prenatal diagnosis of duodenal obstruction is increasing with the routine use of antenatal scans. An altered orientation of superior mesenteric vessels is a very useful screening finding for associated malrotation, which when combined with contributory clinical findings can form an indication for surgery directly without contrast radiography.

Upper gastrointestinal contrast radiography is the gold standard investigation in diagnosing duodenal membrane with a hole. The type of surgery such as the duodenoplasty, duodenoduodenostomy or duodenojejunal anastomosis did not affect the post-op outcome.

Among the associated diseases malrotation, Down's syndrome, genitourinary anomalies, congenital heart disease are commonly found. Anastomotic leak should be kept in mind in every case of duodenal obstruction with post op signs of abdominal distension, bilious NG aspirate and tenderness.

The short-term outcome of patients who underwent uncomplicated Kimura's procedure is good; especially the early intolerance of feeds is less than that reported in literature. However, long-term follow-up is warranted to know the actual incidence of requirement for redo procedure for feed intolerance. The presence of coexisting Down's syndrome and congenital cardiac lesions increases the mortality rate in these children.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the institutional ethics committee

REFERENCES

1. Brinkley MF, Tracy ET, Maxfield CM. Congenital duodenal obstruction: causes and imaging approach. Pediatr Radiol. 2016;46(8):1084-95.
2. Zuccarello B, Spada A, Centorrino A, Turiaco N, Chirico MR, Parisi S. The Modified Kimura's Technique for the Treatment of Duodenal Atresia. Int J Ped. 2009;2009:1-5.
3. Arumugam A, Raja K, Venugopalan M, Chandrasekaran B, Kovanur Sampath K, Muthusamy H, et al. Down syndrome-A narrative review with a focus on anatomical features. Clin Anat. 2016;29(5):568-77.
4. Nijhawan S, Kumpawat S, Mallikarjun P, Bansal RP, Singla D, Ashdhir P, et al. Barium meal follow through with pneumocolon: Screening test for chronic bowel pain. World J Gastroenterol. 2008;14(43):6694-8.
5. Kaldor J. Atresia of the duodenum and duodenal diverticula. Ann surg. 1929;89(1):6-11.

6. Powell DM, Othersen HB, Smith CD. Duodenal obstruction in children: The effect of age on presentation and therapy. J Pediatr Surg. 1989;24:777.
7. Porto SO. Jaundice in congenital duodenal obstruction. Am J Dis Child. 1969;117:684.
8. Willwerth BM, Zollinger RM, Izant RJ. Congenital mesocolic (paraduodenal) hernia: Embryologic basis of repair. Am J Surg. 1974;128:358.
9. YK Sarin. Duodenal webs a case series. J Neonatal Surg. 2012;4:28.
10. Devlin HB. Midgut malrotation causing intestinal obstruction in adult patients. Ann R Coll Surg Engl. 1971;48:227.
11. Orzech N, Navarro OM, Langer JC. Is ultrasonography a good screening test for duodenal obstruction? J Pediatr Surg. 2006;41(5):1005-9.

Cite this article as: Chandrasekaran S, Asokaraju A. Clinical profile and predictors of outcome in congenital duodenal obstruction. Int Surg J 2017;4:2605-11.