International Journal of Contemporary Pediatrics
Bhatta P et al. Int J Contemp Pediatr. 2025 Feb;12(2):312-317
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20250102

Case Series

Assessment of the spectrum of enteric duplication cysts and their varied presentations

Priyamvadha Bhatta*, Neeraj Tuteja

Department Of Pediatric Surgery, SMS Medical College, Jaipur, Rajasthan, India

Received: 30 November 2024
Revised: 07 January 2025
Accepted: 14 January 2025

***Correspondence:**
Dr. Priyamvadha Bhatta,
E-mail: drpriyabhatta@gmail.com

ABSTRACT

Enteric duplication cysts having an incidence of 1 in 4500 live births, are not often diagnosed antenatally. Patients who are eventually diagnosed, have a varying presentation, clinically and demographically, demographically and clinically. Classified as cystic and tubular, with the latter being radiologically difficult to detect on ultrasound. Differential diagnoses of mesenteric cysts and omental cysts. This prospective study was done over a two-year period, where patients who were diagnosed with enteric duplication cysts at Sir Padampat Institute of Neonatology and Child Institute, Jaipur were included. They were monitored from the time of diagnosis, assessing demographic data, preoperative status, surgical management, post operative treatment and follow up after discharge. Total14 patients were included, of which a majority were tubular variants of ileal duplication cyst. 3 patients had foregut duplication cysts (1 gastric, 2 oesophageal). The age ranged from 3 days to 10 years, with the average age of 2.2 years. Each child had a varying presentation, some of them presenting with complications like obstruction and perforation. The incidence was more in male, as compared to females (3.6:1 – male:female). Two children had associated anomalies (one had a congenital diaphragmatic hernia, and one had malrotation of the gut). In one child, a foregut duplication cyst was found to be communicating with a dilated segment of the jejunum, which was resected together. Enteric duplication cysts, can have a myriad of presentations, and other associated congenital anomalies. Further studies are required to identify attributing risk factors, regional incidence along with better awareness for antenatal screening.

Keywords: Enteric duplication cysts, Obstruction, Perforation, Malrotation, Congenital diaphragmatic hernia

INTRODUCTION

Enteric duplication cysts (EDC) are congenital anomalies of the gastrointestinal tract, which can mimic other cysts of the abdominal cavity, such as mesenteric cyst and omental cysts. In literature, an incidence of 1 in 4500 live births has been mentioned.[1] However, in a large population like that of India, this incidence may need to be reassessed. EDC, albeit a rare congenital anomaly, commonly present as ileal duplication cysts. The other locations are mediastinal, colonic, gastric, duodenal, rectal, oesophageal and rarest, cervical. They are classified as tubular or cystic.[2] EDCs also present with other anomalies or complications, which may be more emergent and severe. While EDCs may be detected antenatally, many of them may be detected in the first few years of life. They are believed to occur in the 4th to 8th week of embryonic life. Although the exact aetiology is still to be determined, several theories have been proposed to explain their pathophysiology. Theories such as split notochord theory, luminal recanalization and incomplete twinning are popular hypotheses for the development of EDCs.[3] EDCs are rare congenital anomalies; however, we assessed the patients who were brought to our centre and diagnosed with EDC, either preoperatively or intraoperatively. In our study, we demonstrate the varied presentations of duplication cysts, clinically and demographically. We wanted to

demonstrate the various presentations of this congenital anomaly, and other associated anomalies that may mask a duplication cyst.

CASE SERIES

This prospective study, conducted over a two-year period from September 2022 to October 2024, includes patients who presented to Sir Padampat Institute of Neonatology and Pediatric Health, Jaipur, and were diagnosed to have EDCs. These patients were assessed based on their demographic details, history and clinical presentation, associated anomalies, radiological findings, intraoperative findings and management.

They were also followed up in the postoperative period. The patients who were preoperatively suspected to have EDCs, and were then intraoperatively found to have a different diagnosis were then excluded from the study. Few patients who were initially admitted or worked up for a different diagnosis or congenital anomaly and then intraoperatively detected to have an EDC were included in the study. We summaries all the findings in the Table 1.

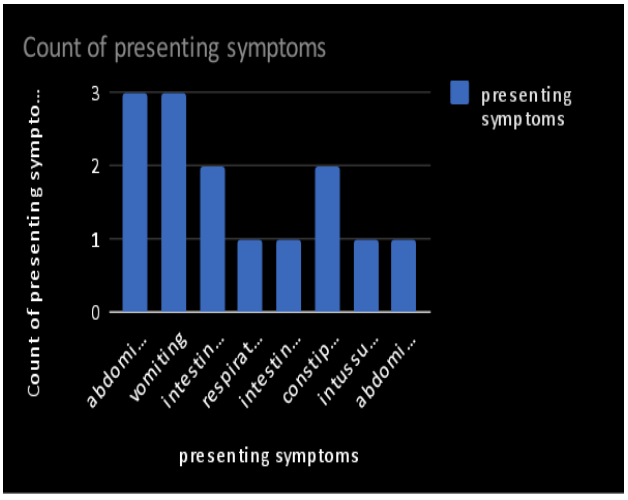

Figure 1: Presenting symptoms.

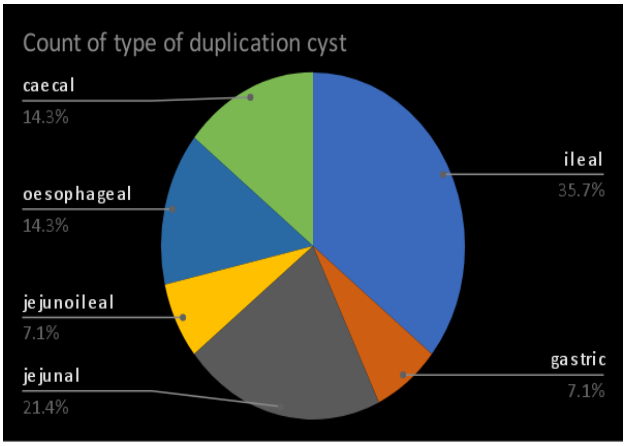

Figure 2: Types of duplication cysts.

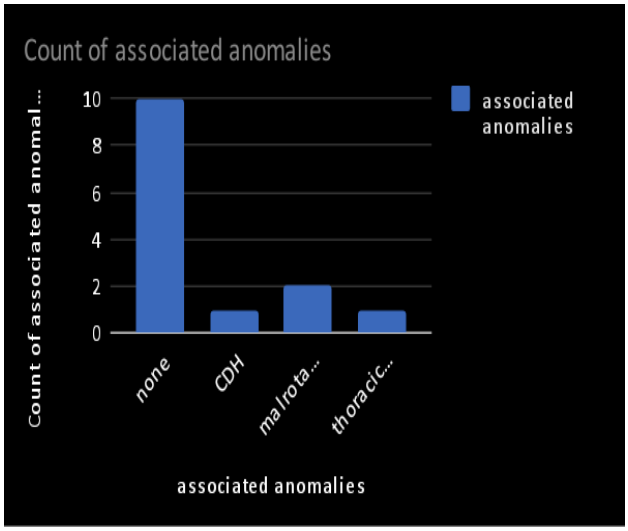

Figure 3: Associated anomalies.

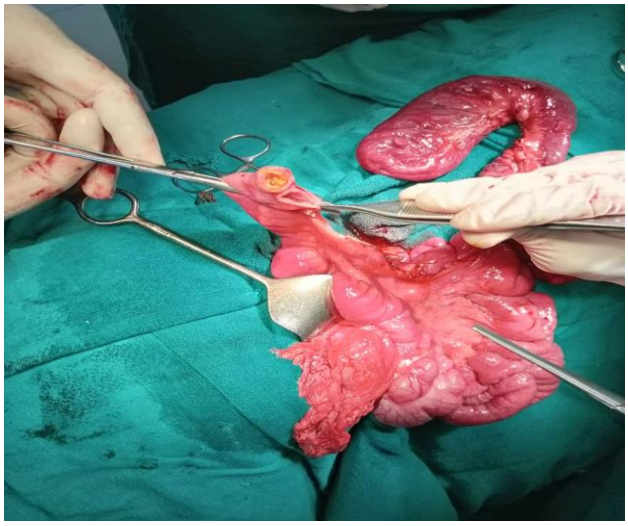

Figure 4: Double lumen with common wall seen of a tubular ileal duplication cyst.

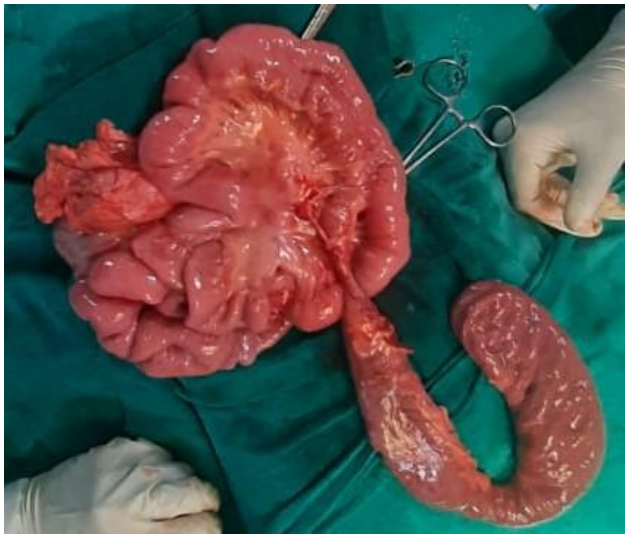

Figure 5: Long tubular ileal duplication cyst.

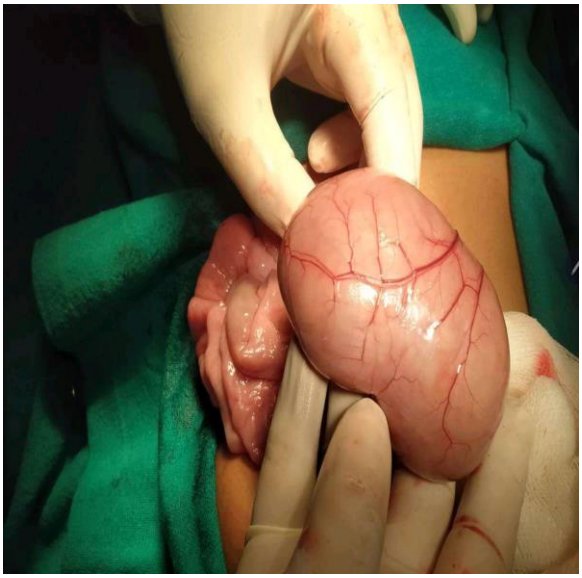

Figure 6: Caecal duplication cyst.

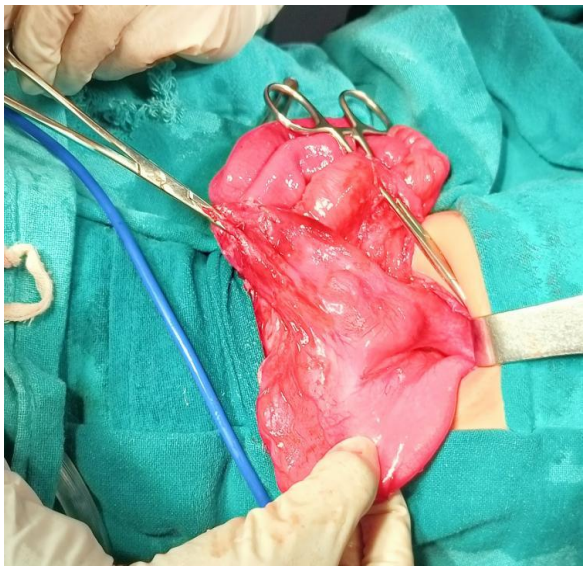

Figure 7: Gastric duplication cyst at the greater curvature of the stomach.

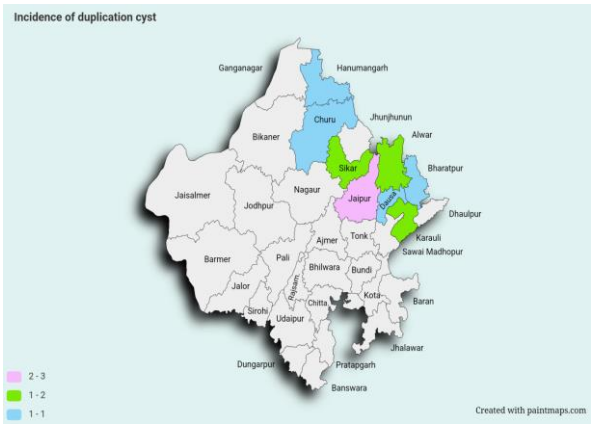

Figure 8: Incidence of duplication cysts in Rajasthan.

The patients that were eventually diagnosed with EDCs presented in a wide age range, beginning from 3 days of age to 10 years of age, with an average age of 2.2 years. The male to female incidence ratio was 3.6:1, with a male preponderance (11 males to 3 females). The most common presenting symptoms for children older than 2 years was found to be vague abdominal pain and recurrent vomiting, while children less than 1 year of age presented with more severe symptoms. (Figure 1). There are 5 patients had ileal duplication cysts, 3 patients had jejunal, 2 patients had esophageal and cecal and one-one patients had gastric and jejuno-ileal duplication cysts (Figure 1). 3 patients presented with complications such as perforation of bowel and intestinal obstruction. 2 patients had associated malrotation of the gut, and one child had a congenital diaphragmatic hernia (Figure 3). Most patients underwent resection and anastomosis of the bowel containing the duplication cyst; however, 2 patients underwent mucosal stripping-one for a jejunoileal duplication cyst, and one for a gastric and duodenal duplication cyst (Figure 4-7). Post operatively, one child had a prolonged ICU stay due to right lung consolidation (CDH with foregut duplication cyst) and one child had a postoperative anastomotic leak, and required re exploration. All patients were discharged, and there were no fatalities.

Table 1: Summary of cases.

S. No	Age/sex	History	Pre-operative diagnosis	Other anomalies	Location of cyst	Treatment	Type of duplication cyst	Tubular/cystic
1	2y/f	Abdominal pain for 6-7 months, intermittent vomiting	Malrotation	None	Ileal duplication cyst	Resection and anastomosis	Ileal	Tubular
2	1y/m	Vomiting for 6-7 days, abdominal distension for 2-3 days	Sealed off perforation	None	Terminal ileal duplication cyst,	Resection and anastomosis	Ileal	Tubular
3	6m/f	Recurrent vomiting for 3-4 months	Duplication cyst/mesenteric cyst	None	Gastric duplication cyst	Deroofing of enteric cyst and stripping	Gastric	Tubular

Continued.

S. No	Age/sex	History	Pre-operative diagnosis	Other anomalies	Location of cyst	Treatment	Type of duplication cyst	Tubular/cystic
4	7m/m	Swelling in the abdomen since birth, abdominal distension and respiratory distress	Intestinal perforation	None	Jejunal duplication cyst with duodenal perforation -	Primary repair of duodenal perforation, excision of jejunal duplication cyst and roux en y gastrojejunostomy	Jejunal	Tubular
5	8d/f	Abdominal distension and bilious vomiting	Intestinal perforation	None	Jejunoileal duplication cyst with inflamed bowel and sealed off perforation of ileum.	Mucosal stripping and proximal ileostomy with distal mucous fistula created	Jejunoileal	Tubular
6	5y/m	On and off and pain for 5 months	Duplication cyst/mesenteric cyst	None	Terminal ileal duplication cyst	Resection and anastomosis	Ileal	Tubular
7	1y6m/m	Breathing difficulties	Congenital diaphragmatic hernia	Congenital diaphragmatic hernia	Oesophageal duplication cyst		Oesophageal	Tubular
8	1y6m/m	On and off abdominal pain and bilious vomiting for 6-7 months	Malrotation with a cystic lesion	Malrotation	Ileal duplication cyst	Resection and anastomosis	Ileal	Cystic
9	3d/m	Abdominal distension and bilious vomiting since birth	Intestinal obstruction	None	Ileal duplication cyst <5 cm from cecum	Resection and anastomosis	Ileal	Cystic
10	8m/m	Abdominal distension on and off since birth, constipation	Redundant colon with mediastinal neurenteric cyst	Thoracic hemi vertebrae	Foregut duplication cyst communicating with a dilated jejunal segment	Resection and anastomosis	Oesophageal	Tubular
11	3y/m	Abdominal pain for 3-4 days, fever	Ileocolic intussusception	None	Caecal duplication cyst	Resection and anastomosis	Caecal	Tubular
12	10y/m	Abdominal pain on and off	Appendicular lump	None	Caecal duplication cyst 5 cm from ic junction	Resection and anastomosis	Caecal	Tubular
13	2y2m/m	Pain abdomen and vomiting for 7 days	Malrotation	None	Jejunal duplication cyst of 10mm,	Resection and anastomosis	Jejunal	Tubular
14	3y/m	Chronic constipation - Hirschprung's disease	Hirschprung's disease	Malrotation	Jejunal duplication cyst 10cm long, 20cm from dj flexure with malrotation	Resection and anastomosis	Jejunal	Tubular

DISCUSSION

The regional incidence was marked on a map of the state, with 3 patients hailing from the city of Jaipur. The incidence of ileal duplication cysts was the highest (35.7%) (Figure 8). Alimentary canal duplication cysts must have three main characteristics**.** An epithelial lining containing the mucosa of the alimentary tract**.** An envelope of smooth muscle**.** The cyst must be closely attached to the GIT by sharing a common wall.[7] On reviewing the existing literature about EDCs, it was noticed that many case reports and few case series were present. The case series that were present, were usually a

small group of cases, or a moderate number that were collected over decades.[2] The term enteric duplication cysts was popularized by Ladd in the 1930's, but was first used by Fitz prior to that.[3] Duplication cysts, while they may also be asymptomatic and present late into adulthood, we found that they can present with complications during the neonatal period and in infancy. In children older than 3 years of age, they presented with vague symptoms. We also found EDCs to be incidentally detected in patients with other congenital anomalies, such as congenital diaphragmatic hernia and malrotation. A case report of a 15-year-old found to have a posterior mediastinal cyst and a large hiatal hernia was managed thoracoscopically for the mediastinal cyst, which was found to be in the left chest cavity, with a type A Bochdalek hernia that was missed on the initial thoracoscopy.[4]

In our study, we had a child who presented with respiratory distress and was found to have a large left diaphragmatic hernia, with right lung consolidation and a foregut duplication cyst. As the need was to resolve the respiratory complaints, the diaphragmatic hernia was tackled first, with the foregut duplication cyst left in situ, to plan for excision at a later date. Bui et al, reported a 2-year-old boy with non-bilious vomiting and abdominal pain, with fusion and segmentation anomalies of the spine, from C5 to T4, along with 4 absent ribs on the left side.[5]

Additionally, on the CT of the chest and abdomen, a possible anterolateral thoracic myelocoele was also seen, with a separate cystic structure also visualized. The cystic lesion extended into the right lower quadrant of the abdomen. Intraoperatively, an abnormal bowel-like cystic structure within the jejunal mesentery extending cephalad was present, crossing the SMA and coeliac axis.

Malrotation of the intestine, with the entire bowel on the right side was seen, with appendix and caecum on the left upper quadrant. The malrotation was corrected, and the cyst was separated from the adjacent bowel wall. On entering the thorax, the extension of the cyst was seen in the posterior mediastinum, and was seen separate from an anterior myelocoele that was at the apex of the left chest. The cystic mass was resected, with repair of the myelocoele done as well as the repair of the diaphragmatic hernia. In our study, a similar patient presented with abdominal distention and constipation. CECT thorax and abdomen was done, which showed a cystic lesion in the mediastinum with hemi vertebral anomalies, suspecting a neuromeric cyst. This child intraoperatively had a mediastinal duplication cyst that extended into the abdominal cavity and ended in a dilated jejunal segment.

A case report by Pai et al, demonstrated a rare case of duplication cyst with midgut volvulus, seen in a 4-day old child. In our series, we had two such patients, who presented after 1 year of age with malrotation. One child was found to have a jejunal duplication cyst, and the other one had an ileal duplication cyst.[6] A Ladd's procedure with appendectomy was done for both our patients, along with resection and anastomosis of the bowel containing the cyst. This was a similar management to that reported by Azzam et al.[8]

At our centre, we encountered two patients with cecal duplication, one of whom presented with intussusception, and the other with a palpable lump in the right iliac fossa. Caecal duplication cysts are a rarer entity of EDCs, accounting for 0.4% of all duplications.[9]

Patients could be misdiagnosed with an appendicular lump, appendicular abscess, or even necrotising enterocolitis. Temiz et al, had done cyst excision with the caecum in 6 of the 7 patients, along with an ileocolostomy, similar to what was performed at our centre for the 2 cecal duplication cysts.[9]

CONCLUSION

EDCs are a rare gastrointestinal anomaly that requires a high index of suspicion when it comes to a preoperative diagnosis. It can present as benign vague abdominal pain that is intermittent, or can present along with bowel perforation, leading to sepsis and shock. They can mimic mesenteric cysts or omental cysts based on their size and location. They can also be associated with other anomalies like malrotation of gut and congenital diaphragmatic hernia. While the combination of EDC with other anomalies is extremely rare, it does prompt further enquiries and studies related to the causality of both occurring in the same patient.

This study has demonstrated a relatively large volume of patients who were diagnosed with EDC during a two-year period, which also prompts more questions about the association of this rare entity and the environmental demography as well. Further studies, classified by region, can aid in determining the association between other anomalies and EDCs.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Sangüesa NC, Llorens SR, Carazo PE, Picó AS, Ibañez PV. Enteric duplication cysts in children: varied presentations, varied imaging findings. Insights into imaging. 2018;9:1097-106.
2. Ildstad ST, Tollerud DJ, Weiss RG, Ryan DP, McGowan MA, Martin LW. Duplications of the alimentary tract. Clinical characteristics, preferred treatment, and associated malformations. Ann Surg. 1988;208(2):184.

3. Sharma S, Yadav AK, Mandal AK, Zaheer S, Yadav DK, Samie A. Enteric duplication cysts in children: a clinicopathological dilemma. JCDR. 2015;9(8):8.
4. Bruckner K, Kurashima M, Blewett C, Miyata S, Herman R. Surgical management of foregut duplication cyst existing with a congenital diaphragmatic hernia: A case report. Int J Surg Case Reports. 2024;124:110369.
5. Bui T, Bankhart MF, Mandell GA, Dickman PS, Bae JO. Thoraco-abdominal enteric duplication cyst in association with neurenteric cyst, axial skeletal anomalies, and malrotation. Radiol Case Rep. 2013;8(1):779.
6. Pai NG, Prabhu S, Prabhakar P, Kumar V. Duplication cyst with midgut volvulus in a neonate: an unusual presentation. BMJ Case Reports CP. 2020;13(8):235526.
7. Sangüesa NC, Llorens SR, Carazo PE, Picó AS, Ibañez PV. Enteric duplication cysts in children: varied presentations, varied imaging findings. Insights into imaging. 2018;9:1097-106.
8. Azzam A, Abdulkarim AN, Shehata AE, Mahran I, Arafa A, Arafat A, et al. A report of two infant cases operated for jejunal duplication cyst associated with malrotation and volvulus. Inter J Surg Case Rep. 2020;67:227-30.
9. Temiz A, Oğuzkurt P, Ezer SS, İnce E, Gezer HÖ, Hiçsönmez A. Different clinical presentations, diagnostic difficulties, and management of cecal duplication. J Pedia Surg. 2013;48(3):550-4.

Cite this article as: Bhatta P, Tuteja N. Assessment of the spectrum of enteric duplication cysts and their varied presentations. Int J Contemp Pediatr 2025;12:312-7.