

Original Research Article

Audit of neonatal congenital anomalies required surgical intervention at tertiary care centre

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ABSTRACT

Background: Nearly 10% of neonatal deaths are due to congenital malformations requiring surgical intervention. Hence our aim is to study spectrum and outcome of the different neonatal congenital anomalies requiring surgical intervention.

Methods: This prospective study was conducted over a period of 2 years. 130 cases which required surgical intervention in neonatal period were included in the study. Plain x-ray abdomen was done in all the cases of our study. Ultrasound scan was done in all the cases to rule out renal and other anomalies. Contrast radiography was also performed in selected cases. All cases underwent their respective operations depending upon the diagnoses. Complication and mortalities during hospital stay were noted.

Results: During the study period total 130 neonates underwent surgical intervention. Out of 130 cases 5(3.84%) neonates had trachea-esophageal fistula, 2(1.53%) had pure esophageal atresia, 5(3.84%) had duodenal atresia, 9(6.92%) had jejunal atresia, 14(10.76%) had ileal atresia, 3 had meconium ileus(2.30%), 9(6.92%) had malrotation, 15(11.5%) had HD and 5(3.84%) had Meckel's diverticulum, 10(7.69%) had Hypertrophic Pyloric Stenosis, 2(1.53%) had gastroschisis, 3(2.30%) had omphalocele, 16(12.30%) had anorectal malformation, 4(3.07%) had Patent Vitello Intestinal Duct, 3(2.30%) had persistent patent urachus, 4(3.07%) had congenital diaphragmatic hernia, 1(0.76%) had Congenital Lobar Emphysema, 4(3.07%) had Neural Tube Defects, 8(6.15%) had Inguinal Hernia, 6(4.61%) had Posterior Urethral Valve and 2(1.53%) had Pelvi-ureteric Junction Obstruction. There were 85 males and 45 females (M: F-2:1). Septicaemia (40%) was most common complication, 21 (16.15%) cases had mortality.

Conclusions: There is lack of awareness regarding dog bite and its management among the rural population.

Keywords: Congenital malformation, Complications, Intestinal atresia, Neonatal mortality, Septicaemia, Surgical intervention

INTRODUCTION

Congenital anomalies are structural or functional anomalies which occur during intrauterine life and can be identified antenatally, at birth or later in life. Congenital anomalies can be caused by single gene defects, chromosomal disorders, multi-factorial inheritance, environmental teratogens and micronutrient deficiencies.^{1,2}

Congenital anomalies account for 11% of neonatal deaths globally and 9% in India. The prevalence of birth anomalies in India is 6-7%.^{3,4}

Birth asphyxia and infections are major contributors to neonatal deaths.⁵ However there are other silent contributors to these deaths which are usually not highlighted. Amongst these, are surgical diseases in the newborn.⁶

Nearly 10% of neonatal deaths are due to congenital malformations requiring surgical intervention.⁷

There has been a tremendous progress in the prenatal diagnosis of Congenital Malformation because of improvements in fetal ultrasound and prenatal genetic testing. This allows parent's the choice of terminating the pregnancy. In the past 2 decades, there has also been a concordant increase in the rate of termination of pregnancy for fetal anomalies.^{8,9}

Early identification of anomalies is important for better outcome. Parent's may feel anxious and guilt on knowing the existence of a congenital anomaly and require sensitive counselling.¹⁰

There have been very few published data detailing the outcome of different neonatal congenital anomalies requiring surgical intervention in India. Hence aim of the study is to determine the spectrum and outcome of the different neonatal congenital anomalies requiring surgical intervention, operated and managed in a tertiary NICU.

METHODS

This prospective study was conducted over a period of 2 years from June 2017 to May 2019. 130 cases which required surgical intervention in neonatal period were included in the study.

Neonates with Multiple anomalies were excluded from the study.

All neonates were admitted, and their dehydration and electrolytes imbalance were corrected. Vitamin K and prophylactic broad-spectrum antibiotics were started.

Plain x-ray abdomen was done in all the cases of our study. Ultrasound scan was done in all the cases to rule out renal and other anomalies. Contrast radiography was also performed in selected cases.

All cases underwent their respective operations depending upon the diagnoses. Complication and mortalities during hospital stay were noted.

SPSS software, Version 18.0 was used for statistical analysis

RESULTS

During the study period total 130 neonates underwent surgical intervention. Out of 130 cases 5(3.84%) neonates had trachea-oesophageal fistula, 2(1.53%) had Pure oesophageal atresia (Figure 1 Chest X ray showing coiling of NG tube with gasless abdomen suggestive of Pure oesophageal atresia), 5(3.84%) had duodenal atresia, 9(6.92%) had jejunal atresia, 14(10.76%) had ileal atresia (Figure 2 Intraoperative image showing Intestinal atresia), 3(2.30%) had meconium ileus.



Figure 1: Chest X ray showing coiling of NG tube with gasless abdomen suggestive of pure esophageal atresia.



Figure 2: Intraoperative image showing intestinal atresia.

(Figure 3 Showing dilated proximal bowel and collapsed distal bowel due to Meconium Ileus), 9(6.92%) had malrotation, 15(11.5%) had Hirschsprung's disease and 5(3.84%) had Meckel's diverticulum, 10(7.69%) had Hypertrophic Pyloric Stenosis (Figure 4 Intraoperative image of pyloromyotomy for Idiopathic Hypertrophic Pyloric Stenosis), 2(1.53%) had Gastrochisis (Figure 5 Showing Gastrochisis), 3(2.30%) had omphalocele, 16(12.30%) had Anorectal malformation, 4(3.07%) had Patent Vitello Intestinal Duct, 3(2.30%) had Persistent patent Urachus, 4(3.07%) had Congenital Diaphragmatic Hernia, 1(0.76%) had Congenital Lobar Emphysema, 4(3.07%) had Neural Tube Defects (Figure 6 Showing Open Neural Tube defect), 8(6.15%) had Inguinal Hernia, 6(4.61%) had Posterior Urethral Valve and 2(1.53%) had Pelvi-ureteric Junction Obstruction. There were 85 males

and 45 females (M:F 2:1). Among males, 18(21%) had intestinal atresia, 2(2.35%) had meconium ileus, 7(8%) had malrotation, 9(10%) had Hirschsprung's Disease, 5(5.88%) had malrotation, and 2(2.35%) had Meckel's diverticulum. Age of presentation varied from 1 day to 24 days. Median weight for patients with intestinal atresia and meconium ileus were 2 Kg (1.3-3 kg), Median weight was 2.5 kg for malrotation and Meckel's diverticulum and for Hirschsprung's disease it was 2.5 kg (1.6-2.9 kg). Intestinal atresia cases comprised of total 28 (21.53%) cases.



Figure 3: Dilated proximal bowel and collapsed distal bowel due to meconium ileus.



Figure 4: Intraoperative image of pyloromyotomy for idiopathic hypertrophic pyloric stenosis.



Figure 5: Gastroschisis.

Among these 5 cases (17.8%) of duodenal atresia , 9 (32.1%) cases of jejunal atresia, 14 (50%) patients of ileal atresia. Type I was the most common type of duodenal atresia. In contrast, type III was the most common type in patients with jejuna and ileal atresia. Out of 5 cases of duodenal atresia, 5 (100%) patients had type I atresia. Out of 14 cases of ileal atresia- 3 (21.4%) had type I, 9 (64.3%) had type III, and 2 (14%) had type IV atresia. Out of 9 cases of jejunal atresia- 7 (77.8%) had type III and 2 (22.2%) had type IV atresia.



Figure 6: Open neural tube defect.

Tracheoesophageal fistula, pure esophageal atresia, Duodenal Atresia, Jejunal Atresia, Ileal Atresia, Meconium ileus, Gastroschisis, omphalocele, anorectal malformation, Congenital Diaphragmatic Hernia cases presented with symptoms within 3 days of life and cases with malrotation, Hirshprung disease, Congenital Hypertrophic pyloric stenosis, Persistant patent Urachus, Inguinal Hernia, Pelviureteric Junction Obstruction presented between 4 days to 28 days of life. Babies with atresia and meconium ileus presented earlier compared to those with Hirschsprung's disease and other causes (Table 1 Age of presentation).

Tracheoesophageal fistula, pure esophageal atresia cases presented with excessive salivation and inability to feed. Duodenal Atresia, Jejunal Atresia, Ileal Atresia, Meconium ileus, Gastroschisis, omphalocele, Congenital Hypertrophic pyloric stenosis cases presented with vomiting. Not passage of stool was presentation in Tracheoesophageal fistula, pure esophageal atresia, Duodenal Atresia, Jejunal Atresia, Ileal Atresia, Meconium ileus, Gastroschisis, omphalocele, anorectal malformation, Hirschsprung disease. Hirschsprung disease. Respiratory distress was main presentation in cases of Tracheoesophageal fistula, pure esophageal atresia, Congenital Diaphragmatic Hernia, Congenital Lobar Emphysema. Bilious vomiting, abdominal distention and failure to pass meconium or stool were the most prominent features of presentation in most cases (Table 2a) .

Neural Tube Defects were presented with open defect and discharge from back, Inguinal hernia presented with swelling, Posterior urethral Valve presented with

difficulty in urination and Pelviureteric Junction Obstruction presented with Antenatal gross hydronephrosis (Table 2b).

Septicaemia (40%) was most common complication, followed by pneumonitis (12.3%), wound infection (4%) and anastomotic leak (2.3%) (Table 3).

Table 1: Age at presentation.

Causes	0-3 days	4-15 days	15-28 days
Tracheo-oesophageal Fistula	4 (80%)	1 (20%)	0
Pure oesophageal atresia	2 (100%)	0	0
Duodenal atresia	4 (80%)	1 (20%)	0
Jejunal atresia	7 (78 %)	2 (22%)	0
Ileal atresia	12 (85%)	2 (15%)	0
Meuconium ileus	3 (100%)	0	0
Malrotation	0	5 (55%)	4 (45%)
Hirshprung disease	0	2 (13%)	13 (87%)
Meckles diverticulum	0	0	5 (100%)
Hypertophic pyloric stenosis	0	0	10 (100%)
Gastrochesis	2 (100%)	0	0
Omphalocele	2 (67%)	1 (33%)	0
Anorectal malformation	16 (100%)	0	0
Patent vitelo intestinal duct	2 (50%)	2 (50%)	0
Persistant patent urachus	0	0	3 (100%)
Congenital diaphragmatic hernia	3 (75%)	1 (25%)	0
Congenital lobar emphysema	0		1 (100%)
Neural tube defects	2 (50%)	2 (50%)	0
Inguinal hernia	0	3 (37%)	5 (63%)
Posterior urethral valve	5 (83.3%)	1 (18%)	0
Pelviureteric junction obstruction	0	2 (100%)	0

Table 2a: Clinical presentation.

Causes	Vomiting/excessive salivation	Not passed stool since birth	Abdominal distention	Respiratory distress	Umbilical discharge
Tracheo-oesophageal fistula	5 (100%)	0	3 (60%)	4 (80%)	0
Pure oesophageal atresia	2 (100%)	2 (100%)	0	1 (50%)	0
Duodenal atresia	5 (100%)	5 (100%)	0	1 (20%)	0
Jejunal atresia	9 (100%)	9 (100%)	4 (44%)	0	0
Ileal atresia	14(100%)	14 (100%)	12 (85%)	2 (14%)	0
Meuconium ileus	3 (100%)	3 (100%)	3 (100%)	0	0
Malrotation	9 (100%)	0	5 (55%)	0	0
Hirshprung disease	8 (53%)	0	15 (100%)	0	0
Meckles diverticulum	5 (100%)	0	4 (80%)	0	0
Hypertophic pyloric stenosis	10(100%)	0	0	0	0
Gastrochesis	2 (100%)	2 (100%)	0	0	0
Omphalocele	1 (33%)	0	0	0	0
Anorectal malformation	2 (12%)	16 (100%)	14 (88%)	0	0
Patent vitelo intestinal duct	1 (25%)	0	0	0	4 (100%)
Persistant patent urachus	0	0	0	0	3 (100%)
Congenital diaphragmatic hernia	0	0	0	4 (100%)	0
Congenital lobar emphysema	0	0	0	1 (100%)	0

One case of jejunal atresia and 2 cases of ileal atresia were re-explored due to anastomotic leak.Surgical intervention was done in all cases of congenital

anomalies,21(16.15%) cases had mortality. Maximum mortality was with Gastrochesis (100%) followed by Neural Tube Defects (75%) , Omphalocele (67%),

Tracheo-oesophageal fistula (60%)No mortality seen in cases of Meconium ileus, Malrotation, Meckels Diverticulum , Congenital Hypertrophic pyloric stenosis,

Anorectal malformation, Patent Vitelo Intestinal Duct , Persistant patent Urachus , Inguinal Hernia, Pelviureteric Junction Obstruction (Table 4).

Table 2b: Clinical presentation.

Causes	Presentation
Neural tube defects	Open defect, Discharge back (4) (100%)
Inguinal hernia	Swelling (8) (100%)
Posterior urethral valve	Difficulty in urination (6) (100%)
Pelviureteric junction obstruction	Antenatal gross hydronephrosis (2) (100%)

Table 3: Complication.

Causes	Number	Septicemia	Wound infection	Anastomotic leak	Pneumonitis
Tracheo-oesophageal fistula	5	4 (80%)	1 (20%)	0	4 (80%)
Pure oesophageal atresia	2	1 (50%)	0		1 (50%)
Duodenal atresia	5	3 (60%)	0		1 (20%)
Jejunal atresia	9	6 (67%)	1 (11%)	1 (11%)	
Ileal atresia	14	9 (64%)	0	2 (14%)	3 (21%)
Meconium ileus	3	2 (67%)	0	0	0
Malrotation	9	4 (44%)	0	0	0
Hirschsprung disease	15	3 (20%)	1 (6.7%)	0	0
Meckles diverticulum	5	2 (40%)	0	0	0
Hypertrophic pyloric stenosis	10	2 (20%)	0	0	0
Gastroschisis	2	2 (100%)	0	0	2 (100%)
Omphalocele	3	2 (67%)	0	0	2 (67%)
Anorectal malformation	16	3 (19%)	0	0	0
Patent vitelo intestinal duct	4	2 (50%)	1 (25%)	0	0
Persistent patent urachus	3	0	1 (33%)	0	0
Congenital diaphragmatic hernia	4	2 (50%)	0	0	2 (50%)
Congenital lobar emphysema	1	1 (100%)	0	0	1 (100%)
Neural tube defects	4	2 (50%)	1 (25%)	0	0
Inguinal hernia	8	0	0	0	0
Posterior urethral valve	6	2 (33%)	0	0	0
Pelviureteric junction obstruction	2	0	0	0	0
Total	130	52 (40%)	6 (4.60%)	3 (2.30%)	16 (12.30%)

DISCUSSION

Several Congenital Anomalies amenable to surgery will, if not recognized and adequately treated, lead to death within a few days after birth. These conditions constitute surgical emergencies of the newborn. High mortality rates due to delayed treatment caused by the paucity of health professionals trained to identify and treat anomalies and by cultural beliefs surrounding anomalies. Surgical congenital malformation requires exposure of every paediatric trainee in recognition of surgical problems in babies in training and referral at the earliest to Paediatric Surgeon. A sophisticated infrastructure is

required for management of surgical congenital malformation to confirm diagnosis and surgical intervention by experienced surgeons followed by dedicated neonatal intensive care for post-operative support. In this study we found that the most common anomalies were intestinal atresia(21.53%) followed by Anorectal malformation (12.30%), followed by Hirschsprung’s disease (11.5%) and that males were affected more than the females. This is comparable to other studies from developing countries.11-17Mortality rate in congenital intestinal atresia in present study was 25% (7 among 28).The mortality associated with intestinal obstruction ranges from 21-45% in developing countries and it is less than 15% in European countries.

Table 4: Surgical intervention and mortality.

Causes	Number (%)	Surgery done	Mortality
Tracheo-oesophageal Fistula	5 (3.8%)	Right Thoracotomy, Fistula ligation and esophagoesophageal anastomosis	3 (60%)
Pure oesophageal atresia	2 (1.5%)	Oesophagostomy and feeding gastrostomy	1 (50%)
Duodenal atresia	5 (3.8%)	(%)Kimura's duodeno-duodenostomy	2 (40%)
Jejunal atresia	9 (6.9%)	Resection and anastomosis	2 (22%)
Ileal atresia	14 (10.77%)	resection and anastomosis	3 (21%)
Meuconium ileus	3 (2.3%)	enterotomy	0
Malrotation	9 (6.9%)	Ladd's procedure	0
Hirschsprung disease	15 (11.5%)	Levelling costomy	1 (6.7%)
Meckles diverticulum	5 (3.8%)	resection and anastomosis	0
Hypertrophic pyloric stenosis	10 (7.7%)	Ramstad's pyloromyotomy	0
Gastroschisis	2 (1.5%)	Abdominal wall repair	2 (100%)
Omphalocele	3 (2.3%)	Abdominal wall repair	2 (67%)
Anorectal malformation	16 (12.30%)	Divided sigmoid colostomy	0
Patent vitelo intestinal duct	4 (3.08%)	Resection and anastomosis	0
Persistent patent urachus	3 (2.3%)	Umbilical exploration and closure of uracus	0
Congenital diaphragmatic hernia	4 (3.08%)	Diaphragmatic repair	2 (50%)
Congenital lobar emphysema	1 (0.77%)	Thoracotomy and lobectomy	0
Neural tube defects	4 (3.08%)	Neural tube repair	3 (75%)
Inguinal hernia	8 (6.15%)	Herniotomy	0
Posterior urethral Valve	6 (4.6%)	Vesicostomy	0
Pelviureteric junction obstruction	2 (1.5%)	Anderson hynes pyeloplasty	0

Survival of neonates shows better outcome with patient's with lower gastrointestinal tract atresia (ARM) than upper intestinal atresia which is comparable with most of the studies.¹⁸

Septicemia and pneumonitis was the most common postoperative complications observed in the present study which was similar to the study done by others.¹⁹

Malrotation of the gut with or without volvulus was diagnosed in 9 cases (6.92%) which is comparable to other studies.¹³⁻¹⁵

The diagnosis of Idiopathic Hypertrophic Pyloric Stenosis (IHPS) was confirmed in 10 cases (7.69%). The diagnosis of IHPS was made with clinical symptoms of non-bilious projectile vomiting after feeds, palpable mass in the upper abdomen, metabolic alkalosis in blood gases and confirmed with ultrasound abdomen. All cases underwent pyloromyotomy with no mortality. The outcome is similar to the other centres.¹²

Anorectal malformation and Hirschsprung's disease cases underwent staged surgeries. First colostomy in neonatal period then definitive surgery later on and results were comparable with other studies.¹¹⁻¹⁷

The mortality due to CDH in other centres of developing countries and Africa was similar to present study (35-50%), The mortality rates in CDH in developed nations like

in European countries like Portuguese (6.4%) and Korea (6.7%), Japan (7.5%) and Saudi Arabia (12.28%).¹¹⁻¹⁷

Outcome of tracheo-esophageal fistula, Gastroschisis, omphalocele, patent vitello-intestinal duct, patent urachus, Neural tube defects and renal anomalies were comparable with other studies.¹¹⁻¹⁷

CONCLUSION

Congenital anomalies are a major cause of neonatal mortality. Early diagnosis and surgical correction of life-threatening congenital malformation will decrease neonatal mortality and improves the chances for survival.

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Ethical approval: The study was approved by the Institutional Ethics Committee

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