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Congenital Digestive Malformation and Associated Anomalies

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Abstract:

Congenital anomalies of the digestive tract are an important part of congenital anomalies as they constitute one of the main causes of mortality in children. We aimed in our study to determine the most common digestive anomaly associated with other organ anomalies. Our study also sought to ascertain the most frequent associated anomaly in our patients and its connection to mortality, the average age of diagnosis, prenatal diagnosis and gestacional age. The study was conducted in two phases and includes a total of 273 children diagnosed with Congenital anomalies of the digestive tract, presented at the Pediatric Intensive Care Unit in UHC "Mother Teresa". The first phase, retrospective, includes 137 patients during the period January 2006 – December 2010. The second phase of the study, prospective, includes 136 patients from January 2011 - March 2015. The most frequent digestive anomaly in patients with associated anomalies was Anal atresia (26.70%), followed by Esophageal atresia (21.70%). Cardiac congenital anomaly was the most frequent founded in our study, 56 cases (20.50%). In our study, Meconium ileus, Intestinal malrotation and Congenital megacolon are the most associated with other anomalies (100%). It is important to identify cases associated to other congenital anomalies because they condition patient's managment and prognosis. Anomalies that are often associated with other defects have shown early clinical symptoms and require more time and effort for correction.

Key words: congenital digestive anomalies, newborn babies, age of diagnosis, associated organ anomalies, cardiac congenital anomalies.

INTRODUCTION

Congenital anomalies are also known as birth defects, congenital disorders or congenital malformations. Congenital anomalies can be defined as structural or functional anomalies (e.g. metabolic disorders) that occur during intrauterine life and can be identified prenatally, at birth or later in life. Congenital anomalies can be caused by single gene defects, chromosomal disorders, multifactorial inheritance, environmental teratogens and micronutrient deficiencies.[1]

The gastrointestinal tract may be subject to a variety of congenital abnormalities (i.e. those present at birth) that arise during embryological development. Specific patterns of malformations of the gastrointestinal tract include abnormal lumenisation (stenoses and atresias), duplications, abnormal rotation and fixation, abdominal defects and a variety of others associated with persistence of embryonic structures (e.g. Meckel's diverticulum), or abnormal formation of specific regions of the gastrointestinal tract (e.g. microgastria) or its cellular components (e.g. nerves in Hirschsprung's disease). These disorders primarily result in symptoms of intestinal obstruction, effects on surrounding structures or of associated anomalies.[2]

They frequently manifest with feeding difficulties, distention, and emesis at birth or within 1 or 2 days.[3] The clinical symptoms varies depending on the pathology, the age of diagnosis and associated anomalies. They may be from mild to severe with, recurrent abdominal pain, intestinal obstruction, dehydration, malnutrition, malabsorption/diarrhea, peritonitis/septic shock, solid food intolerance, common bile duct obstruction, abdominal distention, and failure to thrive.[4] Some congenital gastrointestinal malformations, such as

malrotation, have a very good outcome, whereas others, such as congenital diaphragmatic hernia, have a poor outcome, with a relatively high mortality rate of 10 to 30%.[3]

Early clinical recognition of these disorders is essential to minimise complications and allow the early institution of appropriate therapies.

Although surgery is the commonest intervention early treatment must include adequate resuscitation and stabilisation of the child prior to definitive surgery. Many of these abnormalities are associated with other congenital anomalies or genetic syndromes and disease making the clinical symptoms and the management more complex and difficult.[2] Congenital cardiac malformations are frequently associated with non-cardiac malformations and chromosomal anomalies. Management is therefore influenced by interventional needs for all of the various anomalies.[5]

Through our study we sought to evaluate the connection of congenital anomalies to congenital digestive anomalies in newborns who were hospitalized in our Intensive Unit Care.

MATERIALS AND METHODS

The study was conducted in two phases and includes a total of 273 children diagnosed with Congenital anomalies of the digestive tract, presented at the Pediatric Intensive Care Unit in UHC "Mother Teresa". The first phase of the study is case-control type or otherwise known as retrospective study. It includes 137 patients, who were admitted to the Pediatric Intensive Care Unit during the period January 2006 – December 2010 with the main diagnose "Congenital gastrointestinal anomalities".

The second phase of the study is prospective type and includes cases presented from January 1, 2011 to March 1, 2015, where patients have been followed for at least 1 month.

The second phase of the study, prospective, includes 136 patients.

Statistical analysis:

Continuous data were presented in average value and in standard deviation. Discrete data were presented in absolute value and in percentage. The correlation between the two variables was analyzed by the Kendal's tau correlation coefficient.

Presentation of the data was done through simple and composite tables and graphs of different types.

The statistical analysis was carried out through the statistical package SPPS 19.0 (Statistical Package for Social Sciences Inc., Chicago II USA) and Microsoft Excel.

Significant values of $p \leq 0.01$ were considered.

RESULTS AND DISCUSSION

In our study are included 273 patients with Congenital digestive anomaly as below in Table 1:

Table 1: Congenital digestive anomaly frequency

Congenital digestive anomaly	No.	Percentage
Anal atresia	59	21.60%
Omphalocele	38	13.90%
Intestinal atresia	50	18.30%
Meconuim ileus	3	1.10%
Hirschsprung's disease	8	3.00%
Duodenal atresia	20	7.30%
Esophageal atresia	48	17.60%
Biliary atresia	6	2.20%
Diaphragmatic hernia	26	9.50%
Intestinal malrotation	3	1.10%
Common mesentery	6	2.20%
Congenital megacolon	2	0.70%
Hypertrophic pyloric stenosis	4	1.50%
Total	273	100.00%

The most frequent anomaly in our study is Anal atresia, 59 cases (21.60%), followed by Intestinal atresia, 50 cases (18.30%) and Esophageal atresia, 48 cases (17.60%).

In a total of 273 cases diagnosed with Congenital gastrointestinal anomaly, 120 of them (44.00%) were also associated with another congenital anomaly.(Table 2)

Table 2: Frequency of associated anomalies in our patients

Associated anomaly	No.	Percentage
Not present	153	56.00%
Present	120	44.00%
TOTAL	273	100.00%

From 120 cases presented with another congenital anomaly, the most frequent are Cardiac anomalies, 56 cases (56.00%), followed by Pulmonary anomalies 21 cases (7.70%) and Abdominal anomalies, 15 cases (5.50%), as shown below.(Table 3)

Table 3: Associated anomalies by their frequency

Associated anomalies	No.	Percentage
No associated anomaly	153	56.00%
Cardiac	56	20.50%
Pulmonary	21	7.70%
Abdominal	15	5.50%
Renal	11	4.00%
Plurianomaly	9	3.30%
Cystic fibrosis	6	2.20%
Palatoschisis	1	0.40%
Pes equinovarus	1	0.40%
TOTAL	273	100.00%

The most common Digestive anomaly founded in 120 cases with associated anomaly is Anal atresia (26.70%) followed by Esophageal atresia (21.70%). (Table 4) Congenital cardiac malformations are frequently associated with non-cardiac malformations and chromosomal anomalies. Management is therefore influenced by interventional needs for all of the various anomalies.[5]

Table 4: Frequency of Digestive anomalies in cases with associated anomalies

Congenital digestive anomaly	No.	Percentage
Anal atresia	32	26.70%
Esophageal atresia	26	21.70%
Omphalocele	18	15.00%
Intestinal atresia	18	15.00%
Diaphragmatic hernia	10	8.30%
Duodenal atresia	7	5.80%
Intestinal malrotation	3	2.50%
Meconuim ileus	3	2.50%
Congenital megacolon	2	1.70%
Common mesentery	1	0.80%
Biliary atresia	0	0%
Hirschsprung's disease	0	0.00%
Hypertrophic pyloric stenosis	0	0%
TOTAL	120	100.00%

Meanwhile, in our study, we have reached in conclusion that Meconium ileus, Intestinal malrotation and Congenital megacolon are the first digestive anomalies most associated with another anomalies, positively with 100% of cases. Biliary atresia, Hirschsprung's disease and Hypertrophic pyloric stenosis, in our study were found to be isolated anomalies. (Table 5)

In a retrospective review made in three Medical centers in Wichita, Kan, was found that patients with Anorectal malformations have a high incidence of associated congenital anomalies. Evaluation of the most commonly affected organ systems in these infants is essential because it is these associated anomalies that account for most of the morbidity and mortality that is associated with this condition.[6]

Table 5: Associated anomalies in each digestive anomaly

Congenital digestive anomaly	Associated anomaly		TOTAL
	Present	Not present	
Anal atresia	32	27	59
	54.20%	45.80%	
Duodenal atresia	7	13	20
	25.00%	65.00%	
Esophageal atresia	26	22	48
	54.20%	45.80%	

	18	32	50
Intestinal atresia	36.00%	64.00%	
	0	6	6
Biliary atresia	0.00%	100.00%	
	10	16	26
Diaphragmatic hernia	38.50%	64.50%	
	3	0	3
Meconuim ileus	100.00%	0.00%	
	3	0	3
Intestinal malrotation	100.00%	0.00%	
	2	0	2
Congenital megacolon	100.00%	0.00%	
	1	5	6
Common mesentery	16.70%	83.30%	
	0	8	8
Hirschsprung's disease	0.00%	100.00%	
	18	20	38
Omphalocele	47.40%	52.60%	
	0	4	4
Hypertrophic pyloric stenosis	0.00%	100.00%	
	120	153	273
TOTAL	100.00%	100.00%	100.00%

In our study was found a significative relation ($p<0.01$) between gestational age and presence of associated anomalies in infants diagnosed with Congenital digestive anomaly. In our preterm babies, most patients had another associated anomaly (57.60%), meanwhile most of term babies in our study (60.50%) had isolated Congenial digestive anomalies. This shows the significance of presence of another anomaly in the progress of pregnancy. (Table 6)

Table 6: Connection of with gestational age

Associated anomalies	**Preterm**	**Term**	**Postterm**
	38	79	3
Present	57.60%	39.50%	42.90%
	28	121	4
Not present	42.40%	60.50%	57.10%
	66	200	7
TOTAL	100.00%	100.00%	100.00%

Another significant connection ($p< 0.01$) was found between the presence of associated anomalies and the diagnostic age of

patients with congental digestive anomalies. The more associated anomalies, the more simptoms are presented, and this is reflected in the age of diagnosis. In cases with another anomaly the average age of diagnosis was about two times smaller than in cases with isolated congenital digestive anomaly.(Table 7)

Table 7: Connection with average diagnostic age

Associated anomalies	Avarage diagnostic age
Present	2.02
Not present	5.65

Between presence of associated anomalies in patients with digestive anomaly and prenatal diagnosis, mortality, presence of complications and average weight was not found a significant correlation ($p > 0.01$).

The prognosis for infants with Congenital digestive anomalies is highly dependent on the presence of associated malformations or karyotype anomalies. Surgery can be performed with low mortality. Morbidity is high for a small group of infants, but the majority of survivors have an uncomplicated surgical course.[7]

CONCLUSION

This research has shown the importance of presence of another anomaly in babies with Congenital digestive anomaly. Babies with more than one congenital anomaly influence on gestational age and average age of diagnostication. Coexistence with multiple anomalies makes the clinic presentation more severe and more complex to achieve a better management. Identification of these patients plays a fundamental role in management and prognosis of patients diagnosed with Congenital digestive anomalies.

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