

Birth Defects: A Hospital Based Study in Chittagong, Bangladesh

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Abstract

Introduction: Birth defects are one of the most common causes of disability in developed and developing countries. Birth defects are structural or functional anomalies, including metabolic disorders, which are present at the time of birth. Although the worldwide incidence of birth defects is estimated at 3-7%, the rate varies widely between countries. In this paper, we categorized the birth defects most commonly seen in Bangladesh and their outcome. We hope to use this information to guide the strategies used to manage these problems. **Materials and Methods:** This hospital based, descriptive study was done in the Department of Pediatric Surgery in Chittagong Medical College and Hospital from January 2008 to December 2012. Records of all patients with birth defects in our hospital and departmental database were reviewed. Types of birth defects, age, sex, surgery performed and outcome were analyzed in patients admitted during this period. Data were analyzed by SPSS 17. Statistical analysis was performed by the chi-square test and mean and percentage values were calculated. $P < 0.05$ was considered to be statistically significant. **Results:** During this five year period, there were 5661 patients of birth defects admitted in this department. Of these, 5156 had a single congenital anomaly and 505 had multiple congenital anomalies. The male to female ratio was 2.1: 1. Birth defects comprised 44.61% of all Pediatric surgical admissions and 0.90% of total hospital admissions. The gastrointestinal system was the most common organ system involved, followed by the genitourinary system. Inguinal hernias were the most common gastrointestinal abnormality and hypospadias were the most common genitourinary case. A total of 3921 operations were performed for birth defects, representing 69.26% of total admission for birth defects. Most operations were done for gastrointestinal and genito-urinary defects. Inguinal herniotomy was the most commonly performed operation followed by laparotomy for various indications. There were 225 deaths for birth defects. While this is only 3.97% of all admissions for birth defects, this represents 51.49% of all (pediatric) deaths during the study period. The most common cause of death was anorectal malformations followed by gastroschisis. **Conclusion:** The study helps assess the burden of birth defects in the Bangladeshi society. There is a need for more extensive, nationwide screening studies to determine the birth prevalence, types and distribution of birth defects in Bangladesh.

Key words: Birth defect; Sex; Systemic distribution; Operations; Outcome.

INTRODUCTION

Birth defects have emerged as the major childhood health problem. Birth defects affect approximately 1 in 33 infants and are the fourth most common cause of neonatal deaths. The disease burden results in about 270 000 neonatal deaths and about 3.2 million birth defect-related disabilities every year¹. A birth defect is any alteration of a normal anatomic structure that is present at birth and has cosmetic, medical or surgical significance.

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It can be narrowly defined in terms of a malformation, an abnormality of physical structure or form usually found at birth or during the first few weeks of life; or defined more widely to include functional disturbances – any irreversible condition existing in a child before birth in which there is sufficient deviation in the usual number, size, shape, location or inherent character of any part, organ, cell or cell constituent to warrant its designation as abnormal²⁻³. The WHO defines a birth defect (also referred to as a congenital anomaly) as a structural or functional anomaly, including metabolic disorders, which are present at the time of birth¹.

The etiology of the malformation can be divided into genetic (multifactorial, single gene, or chromosomal) factors, environmental factors, teratogens and unknown etiologies. Teratogens include maternal conditions such as alcoholism, diabetes, endocrinopathy, phenylketonuria, nutritional deficiency and infections; mechanical problems; chemical agents; drugs; radiation; hyperthermia, etc⁴⁻⁷. Approximately 50% of major anomalies have no recognized etiology and most have multi-factorial inheritance. It is estimated that 94% of serious birth defects occur in middle- and low-income countries, where mothers are more susceptible to macro- and micronutrient malnutrition and have increased exposure to perinatal infection and increased alcohol use during pregnancy. Advanced maternal age also increases the risk of chromosomal abnormalities such as Down syndrome⁴⁻⁸.

Although the worldwide incidence of birth defects is estimated at 3-7%, the rate varies widely between countries⁹. It is reported to be 1.07% in Japan, 1.49% in South Africa, 2% in England, 2-3% in USA and 4.3% in Taiwan. In India, the overall incidence of birth defects ranges from 0.3% to 3.6%. These variations were suggested to be due to social, racial, ecological, and economical influences⁴.

In 2010, the World Health Assembly adopted a resolution to promote primary prevention and the health of children with birth defects. To achieve this aim, the assembly called for the development of registration and surveillance systems, with supplemental goals of developing body of expertise, strengthening research on aetiology, diagnosis and prevention and promoting international cooperation¹. However, despite valiant efforts, many developing countries have no record system for birth defects and data on birth defects from population-based studies originating from developing countries are largely lacking.

Bangladesh is a densely populated country with a high growth rate of 1.37 % a year¹⁰. Many babies are born without any antenatal evaluation and a significant amount of deliveries occur outside the hospital setting. Due to the lack of prenatal screening, maternal awareness of the problem and limited parental knowledge of resources available to them, babies with congenital anomalies never receive adequate medical follow up. If proper care can be given, the morbidity and mortality of birth defects can be reduced. Presently, there is no data available on the incidence of birth defects in Bangladesh or their predisposing factors. This study will attempt to categorize birth defects most commonly seen in Bangladesh with the hope that this information can then be used to better prevent and manage birth defects on the country.

MATERIALS & METHODS

This hospital based, descriptive, retrospective study was done in the Department of Pediatric Surgery, Chittagong Medical College Hospital (CMCH) from January 2008 to December 2012. CMCH is the largest hospital with pediatric surgical facilities in the Chittagong district of Bangladesh. The hospital serves a catchment area with population of approximately 30 million people¹¹. Records of all patients with birth defects in hospital and departmental database were reviewed. The types of birth defect, age, sex, surgery performed and outcome were assessed in patients admitted during this period. Comparisons were made between the total pediatric surgical admissions and total hospital admissions during this period. Stillborn babies with birth defects were excluded from this study. Data were analyzed by SPSS 17. Statistical analysis was performed by the chi-square test; mean and percentage values were calculated. $P < 0.05$ was considered to be statistically significant.

RESULTS

During this five year period, 5661 patients with birth defects were admitted to the Pediatric Surgery Department of CMCH. Of these, 5156 had single congenital anomaly and 505 had multiple congenital anomalies. Birth defects comprised 44.61% of all pediatric surgical admissions and 0.90% of total hospital admissions during this time period. Table-1 shows the percentage of pediatric surgical and total hospital admissions for each year of our study. Males had a higher frequency of congenital anomalies when compared to female (3837 vs. 1761) with a male to female ratio of 2.1: 1. Table-2 shows the gender distribution of birth defects and other pediatric surgical admissions.

Table 1 : Yearly Percentage of Pediatric Surgical and Total Hospital Admissions

Year	Number of Birth Defects	Number of Pediatric Surgical Admissions	Percent of Pediatric Surgical Admissions	Total Hospital Admissions	Percent of Total Hospital Admissions
2008	927	2012	46.07	111753	0.83
2009	1298	2162	60.04	114830	1.13
2010	1143	2470	46.28	130394	0.88
2011	1164	2985	38.99	139424	0.83
2012	1129	3060	36.90	132697	0.85
Total	5661	12689	44.61	629098	0.90

Table 2 : Birth defects by sex

	Number of Pediatric Surgical admissions	Number of birth defects	Percentage
Male	8306	3837	46.20
Female	4320	1761	40.76
Ambiguous genitalia	63	63	100
Total patients	12689	5661	20.18

$P < 0.05$

The gastrointestinal system was the most common organ system involved, followed by the genitourinary system. Inguinal hernias were the most common gastrointestinal abnormality and hypospadias were the most common genitourinary case. Table-3 shows the relative distribution of birth defects by organ systems.

Table 3 : Systemic distribution of birth defects

Gastrointestinal		Genitourinary		Vascular	
Inguinal hernia	1121	Hypospadias	975	Haemangioma	251
ARM	460	PUV	186	Thalassemia with splenomegaly	65
Malrotation	86	UDT	169	Cystic hygroma	53
Intestinal Atresia	72	Phimosis	139	Vascular malformation	29
Omphalocele	68	PUJO	108	Lymphangiohaemangioma	9
IHPS	64	Ambiguous genitalia	63	Hemophilia	4
Gastroschisis	41	Ectopiavesicæ	30	Macrostoma	3
Umbilical hernia	36	Neurogenic bladder	16		
Biliary atresia	24	Ectopic ureter	14	Orofacial	440
Choledocal Cyst	22	Epispadias	13	Tongue tie	144
Diaphragmatic hernia	15	Mega meatus intact prepuce	7	Cleft lip	98
Ventral hernia	13	Torsion of penis	6	Cleft lip with palate	94
Patent VID	11	Urogenital sinus anomaly	6	Cleft palate	80
Eventration of diaphragm	7	Duplex urinary system	5	Preauricular skin tag	24
Meckel's diverticulum	7	Vaginal tag	4		
midgut Volvulus	2	Ectopic kidney	3	Head and neck	51
Para umbilical hernia	2	Horse shoe kidney	3	Meningocele	18
Epigastric hernia	1	Renal agenesis	2	Myelomeningocele	11
Antral web	1	Ectopic testis	2	Branchial fistula	7
Meconium plug	3	Polycystic kidney disease	2	Enancephaly	4
Meconium ileus	3	Ectopic penis	1	Thyroglossal fistula	4
Duplication cyst	2	Patent Urachus	2	Lipomeningocele	3
		Cobb's collar	2	Thyroglossal Cyst	3
Orthopedic	61	Uretrocele	3	Microcephaly	1
Syndactyly	32	urethral duplication	1		
Polydactyly	22	Torsion of penis	3	Skin and soft tissue	93
club foot	6	VUJO	9	Dermoid	86
cong.Hip dislocation	1	Micropenis	5	Sacrocoxygealteratoma	4
				Amniotic band	3

Age of presentation varied among defects, with the youngest infants presenting with gastroschisis and omphaloceles and with oldest with hypospadias and thalassemia with splenomegaly. Figure-1 shows the age range with mean age at presentation and Table-4 shows the mean age at presentation of birth defects at different age groups.

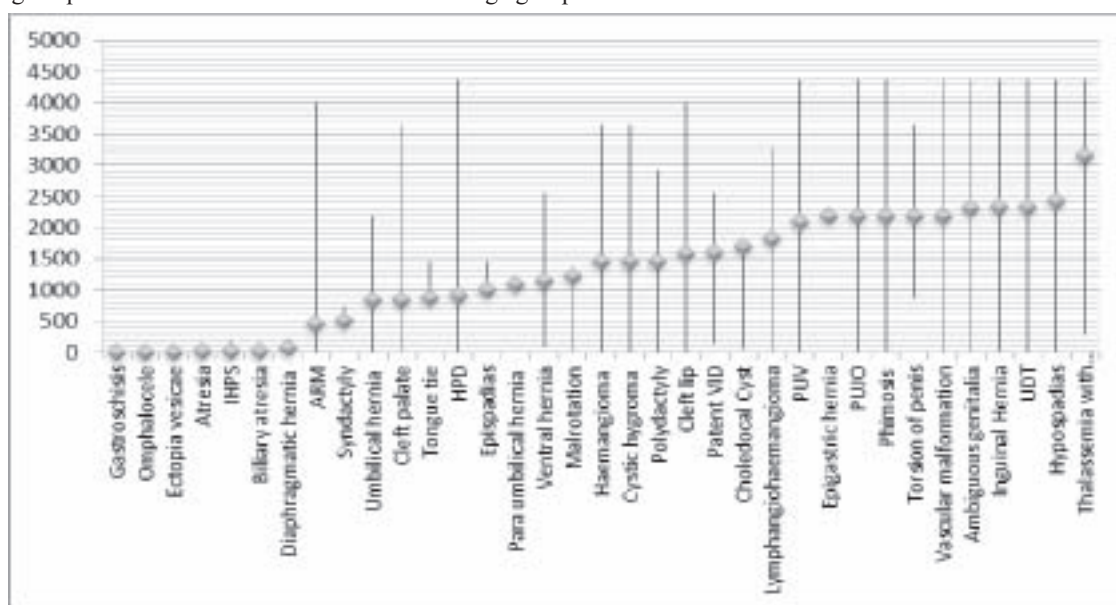


Figure 1 : Age range with mean age at presentation (Days)

Table 4 : Mean age at presentation in different age groups

Neonate	28 D-2 yr	2-4 yrs	4-6 yrs	6-8 yrs
Gastroschisis	Diaphragmatic hernia	Umbilical hernia	Cleft lip	DSD
Omphalocele	ARM	Cleft palate	Patent VID	Inguinal Hernia
Ectopiavesicae	Syndactyly	Tongue tie	Choledocal Cyst	UDT
Intestinal Atresia		Para umbilical hernia	Lymphangiohae	Thalassemia with splenomegaly
IHPS		Epispadias	PUV	Hyospadias
Biliary atresia		Hirschsprung's disease	Epigastric hernia	
		Ventral hernia	PUJO	
		Malrotation	Phimosi	
		Haemangioma	Torsion of penis	
		Cystic hygroma	Vascular malformation	
		Polydactyly		

A total of 3921 operations were done for birth defects, representing 69.26% of total admissions for birth defects and 13.98% of total pediatric surgical admission. These operations excluded mini OT procedures like repair of minor cuts, incision and drainage of small abscesses. Table-5 shows the percentage of operations for birth defects and other causes.

Table 6 : Operations done for birth defects

Gastrointestinal		Urinary		Vascular	
Herniotomy	763	Cystoscopic fulguration	240	Haemangioma Excision	53
Laparotomy	226	Urethroplasty	236	Splenectomy	46
Colostomy	168	Cystoscopic examination	160	Cauterization	41
Colostomy Closer	145	Orchidectomy	130	Sclerotherapy Hemangioma	37
Trans anal pull through	126	A-H pyloplasty	50	Sclerotherapy Cystic hygroma	33
Anoplasty	106	Orthoplasty	36	Hemangioma cauterization	26
Secondary closure	102	Pyeloplasty	33	Sclerotherapy	12
Pyloromyotomy for IHPS	77	Repair of Urethral Fistula	29	Radial ligation of Hemangioma	6
Ileostomy	69	Cutaneous Vesicostomy	19	Lymphangiohemangioma	2
PSARP	48	Meatotomy	18	Diathermy cauterization	1
Correction of malrotation	37	Ureteric reimplantation	14	Ligature of bleeding hemangioma	1
Ileostomy closure	33	Repair of bladder exstrophy	13		
Mass closure of burst abdomen	32	Suprapubiccystostomy	12	Others	
Release of congenital bands	31	Nephrostomy	11	Excision	283
Repair of umbilical hernia	26	Ureteric reimplantation	7	Others	159
Redo Anoplasty	25	Correction of Torsion penis	5	Laparoscopy	122
Others	176	Orchiopexy	4	EUA	30
		Others	30	Diagnistic laparoscopy	7
Orthopedic		Head & Neck		Orofacial	
Release of Syndactyly	9	Branchial sinus	2	Release of Tongue Tie	125
Polydactyly Excision	2	Branchial Fistula	1	Cheiloplasty	25
Skin and soft tissue		Duplication of external auditory canal		Pre auricular skin tag	21
Ganglion	8	Preauricular sinus	1	Palatoplasty	17
Epithelial tag	2				
Release of Amniotic band	2				

There were 225 deaths for birth defects which is 51.49% of all Pediatric Surgical deaths during the study period, 3.97% of all admissions for birth defects, and 1.77% of all Pediatric surgical admissions. Out of these, neonatal deaths were 151 (67.11%), infant mortality was 42 (18.67%) and child mortality was 32(14.22%).

Most common cause of death was ARM followed by Gastroschisis. Table-7 shows the systemic distribution of the deaths.

Table 5 : Percentage of operations for birth defects and other causes

Year	Total operations	Operation for birth defects	Percent of operations
2008	1171	630	53.8
2009	1611	853	52.95
2010	1456	836	57.42
2011	1567	853	54.44
2012	1483	749	50.51
Total	7288	3921	53.8

Most operations were done for gastrointestinal and genitourinary defects. Inguinal herniotomy was the most commonly performed operation followed by laparotomy. Table-6 shows the systemic distribution of all operations.

Table 7 : Systemic distributions of Deaths

Gastrointestinal	187	Urinary	27
ARM	46	PUV	23
Gastroschisis	30	PBS	1
Intestinal atresia	28	PKD	1
Neonatal intestinal obstruction	25	PUJO	2
HPD	17		
Omphalocele	16	Vascular	5
CDH	7	Lymphangioma	3
Meconium Ileus	5	Hemangioma	1
Biliary atresia	3	Vascular malformation	1
IHPS	3		
TEF	3	Others	6
Choledochal cyst	1	Multiple birth defects	3
Congenital bands	1	Conjoint twin	2
Malrotation	1	Thorachoschisis	1
Midgut volvulus	1		

DISCUSSION

In the present descriptive study of birth defects in the Chittagong district of Bangladesh, birth defects were 0.9% of total hospital admissions, similar to rates found on incidence studies. Birth defects comprised about 45% of all pediatric surgical admissions. We found a higher occurrence of birth defects in males than females, a finding that is in contrast to another study in Iran who reported no sex difference¹². Birth defects occurred twice in male than in female. Many studies have documented male preponderance but Parmar et al noted a female preponderance⁹. With regard to pattern of birth defects in the study, the most common system involved was gastrointestinal (45%) followed by genitourinary (31.43%). This finding is consistent with reports from Asam by Dutta et al¹². However, it seems that this is not a universal trend. A study from Gujrat by Parmar et al reported that the central nervous system (65%) was the most system followed by gastrointestinal tract (14%) [9] and reports from Iran, Bahrain, Beirut and Maharashtra found the musculoskeletal system to be the most common system involved¹³⁻¹⁵.

The most common gastrointestinal disease was inguinal hernia, which is different from reports in Iran, Asam and Jammu, India where ARM and Hirschsprung's disease were the most common. The findings that hypospadias was the most common genitourinary disease is consistent with reports from Iran and Jammu and different from Asam. The most common orthopedic disease was Syndactyly, which is different from reports from Jammu and Asam (club foot) and different from Iran (polydactyly). The most common vascular disease was hemangioma, and the most common oro-facial disease was tongue tie, which is different from other studies (cleft lip and palate)^{7,9,12-13}.

The most common neurological disease was meningomyelocele, which is consistent with reports from Jammu but different from Iran (hydrocephalus) and Gujrat (anencephaly) which included still born babies. The orthopedic and neurosurgical cases are dealt with by separate departments and their data were not included in this study. We only recorded the cases which initially came to us and were managed by our department or referred to respective discipline.

Mean age at presentation showed 4 groups when plotted on a graph with group 1 presenting in the neonatal period, group 2 presenting around 500 days (1.4 yrs), group 3 presenting around 1200 days (3.3 yrs) and group 4 around 2000 days (5.5 yrs). Admissions and operations for birth defects varied among years which did not show any linear increase as shown by another study. Operations for birth defect made up more than half (53%) of all operations done during the study period. Prevention of birth defects will reduce a huge operative burden of birth defects. Although CDC reported that most birth defects are found within the first year of life, in this study only 1/4th patients presented before their first year of life and mostly with more serious defects¹⁶. Birth defects were a major cause (52%) of deaths during the study period. About 4% of patients admitted for birth defects died of their disease. Neonatal deaths were much higher, representing 67% of cases that died of a birth defect. The reason for this may be because younger infants presented with more serious diseases and CMCH currently lacks a neonatal intensive care unit and other supports for proper neonatal care. Unfortunately, CMCH is representative of most hospitals in the country where there are minimal systems or supports designed for early life intervention and care. Most common cause of death was ARM followed by Gastroschisis, the outcome of these defects is more favorable in developed countries.

Every 4.5 minutes, a baby is born with a birth defect.¹⁶ WHO is working with the US Centers for Disease Control and Prevention's (CDC) National Center on Birth Defects and Developmental Disabilities and other partners for the prevention of birth defects. The Global Strategy for Women's and Children's Health, launched in 2010 by the UN in collaboration with leaders from governments and other organizations like WHO and UNICEF is crucial to be implemented to improve neonatal and child health.

The International Clearinghouse for Birth Defects Surveillance and Research, which is a voluntary non-profit international organization in official relations with WHO, conducts birth defect surveillance and research programs from around the world in order to investigate and prevent birth defects. Prevention of birth defects will obviously reduce a huge burden on surgical care and ensure better health for the younger generation.

CONCLUSION

The study helps define the burden of birth defects in the Chittagong district of Bangladesh. There is a need for more extensive, nationwide screening studies to determine the birth prevalence, types and distribution of birth defects in Bangladesh.

DISCLOSURE

All the authors declared no competing interest.

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