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A RETROSPECTIVE ANALYSIS OF THE PATTERN OF SURGICAL CONGENITAL MALFORMATIONS IN NORTH EASTERN INDIA

Paediatric Surgery

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ABSTRACT

Background: Congenital anomalies (CAs) originate during intrauterine life and interfere with the body functions. CAs are important causes of childhood mortality, chronic illness, and disability.
Aim: The study aimed to understand the pattern of surgical congenital malformations in paediatric patients of Tomo Riba Institute of Health and Medical Sciences (TRIHMS) hospital and other nearby hospitals for twelve consecutive years (2007–2019).
Methods: This is a retrospective study of 1459 patients (age range 0-14 years) operated at TRIHMS and nearby hospitals of Naharlagun. We collected the data from Operation theatre registers and personal records of operating surgeons from the year 2007 to 2019.
Results: The study revealed that congenital anomalies of the gastrointestinal (48.8%), genitourinary system (45.4%), and musculoskeletal system (1.9%) were the most common types of CAs. Males were more affected with CAs than females (73.8% versus 26.2%).
Conclusion: Congenital malformations of the gastro-intestinal and genitourinary systems were the most common types of CAs in the hospitals. To prevent CAs, there is a need to restrict the prescription of medications that may have a teratogenic effect.

KEYWORDS

Congenital; Malformation; Pattern; Paediatric surgery

1.INTRODUCTION:

Congenital malformations are a global healthcare problem. They are important causes of childhood mortality, chronic illness, and disability. The World Health Organization (WHO) estimated that annually, 303,000 newborns die within the first 4 weeks of birth worldwide due to congenital malformations. As per the WHO, congenital malformations are structural, functional, or metabolic anomalies that originate during intrauterine life and interfere with body functions.[1] These malformations happen either due to defective embryogenesis or intrinsic abnormalities during intrauterine development.[1,2]

Classification of CAs is done according to severity into major and minor anomalies.[1,3] CAs are classified into three groups of severity viz minor, severe, and lethal anomalies. Major anomalies are severe and lethal.[1, 4] However, the international classification of diseases classified CAs according to the affected body system.[5] Congenital anomalies are a major cause of morbidity and mortality among infants and children under 5.[6]An estimated 295 000 newborns die within 28 days of birth every year due to congenital anomalies worldwide.[7] About 95% of the children who died from CAs were from middle- and low-income countries.[1, 8] Congenital anomalies are associated with 8–15% of perinatal deaths and 13–16% of neonatal deaths in this country.[9] If not managed properly, congenital anomalies can lead to long-term physical, mental, visual, and auditory disabilities.[1, 10] The exact cause of CAs is unknown in about 40–60% of cases. The etiology of CAs can be genetic, infectious, nutritional, or environmental factors.[7]

Prevention during pregnancy needs risk identification and management.[6] Accurate and timely national estimates of the prevalence of congenital malformations are needed for monitoring trends, assessing preventive efforts, determining service planning, and understanding burden of the disease. The risk factors of occurrence of CAs include genetic disorders, socioeconomic and demographic factors, nutritional factors like maternal obesity, infections during pregnancy, taking certain drugs, ionizing radiation, chemicals, and air pollution. Pregnancy-associated conditions such as insulin-dependent diabetes, hypertension during pregnancy such as antepartum hemorrhage were also found to be associated with more CAs.[1]

According to international data about 2-3% of babies are born with significant congenital birth defects.[4] Limited data is available on the different patterns of congenital anomalies from Arunachal Pradesh. The aim of this study was to estimate the pattern of surgical congenital malformations in Tomo Riba Institute of Health and Medical Sciences (TRIHMS) and other hospitals at Naharlagun town, Arunachal Pradesh for twelve consecutive years (2007–2019).

2. MATERIALS AND METHODS:

2.1 METHODS:

This is a retrospective analysis of patients with congenital anomalies treated at Tomo Riba Institute of Health and Medical Sciences (TRIHMS) and nearby hospitals of Naharlagun. We collected the data from Operation theatre registers and hospital records from the year 2007 to 2019.

2.2. INCLUSION CRITERIA:

We included all pediatric patients below 14 years with surgical malformations operated or examined at the department of surgery TRIHMS Naharlagun or nearby hospitals from June 2007 to December 2019.

2.3 EXCLUSION CRITERIA:

We excluded patients operated in hospitals other than Naharlagun city and patients older than 14 years and all patients managed by other department of TRIHMS.

2.4 OUTCOME MEASURES:

Distribution of congenital anomalies based on organ systems and type of interventions. Differences in different organ system involvement between male and female genders. Demographic data (age, sex, profession, and comorbidities) were also recorded.

2.5 STATISTICALANALYSIS:

The collected data were revised, coded, and analyzed using SPSS version 20 (IBM, USA). Abnormalities were classified based on the primary abnormality as defined under ICD 10. Descriptive statistics were calculated. Analytical statistics such as chi-square and odds ratio were done. Differences at p-value < 0.05 were considered statistically significant.

3. RESULTS

A total number of 1459 cases of CAs were recorded in TRIHMS Hospital, Arunachal Pradesh, during 2007–2019. The majority of the included patients were male (73.9%) while 26.1% were female. 47% of the patients were below 2 years of age, 46.8% were between 2 to 12 years while the rest 6.2% were above 12 years. (Table 1) As regards the different types of anomalies throughout the studied years, gastrointestinal system anomalies were the most frequent type of CAs, constituting 48.8.0%, followed by the genitourinary system (45.4%), musculoskeletal anomalies (1.9%), circulatory system anomalies (1.8%), central nervous system disorders (1.6%), skin anomalies (0.4%), and endocrine disorders (0.1%). Gastrointestinal anomalies were significantly more common in females than males, while genitourinary anomalies were significantly more common in males than females.(Figure 1)(Table 2) Different surgical procedures conducted in the study cohort included cleft lip and palate repair (522), herniotomy(451), excision(86), orchiopexy(41), colostomy(39), anal procedures(37), VP shunting, removal and revision(24), Swenson Pull-Through Procedure(19), Electrocauterization(10) and other miscellaneous surgeries(224).; 6 patients were managed conservatively. (Table 3)

There were few complications during treatment of the anomalies. Fourteen patients had recurrence post-surgery, one patient had bladder injury, five patients had bad recovery, five patients failed treatment, one patient had CSF leak, and one patient had retention of urine, one patient had wound infection and two had wound dehiscence. Two patients died (one with anorectal malformation (ARM), bilateral renal agenesis, and another with choanal atresia, renal agenesis, ARM, bilateral undescended testis, genital hypertrophy). Both of them were managed conservatively. Another patient with patent ductus arteriosus (PDA) died on operation table due to cardiac arrest. (Table 4) Five more patients were lost to follow up.

Table 1: Demographic characteristics of children with CAs reported in the Tomo Riba Institute of Health and Medical Sciences (TRIHMS), Arunachal Pradesh 2007–2019

Demographic Characteristics	No. of patients	Percentage
Age		
Infant (0-2 years)	686	47%
Childhood(2-12years)	684	46.9%
Adolescence>12 years	89	6.1%
Sex		
Male	1078	73.9%
Female	381	26.1%
Total	1459	

Table 2: Frequency of different types of CAs by gender throughout the studied years in June 2007 to December 2019

	Male		Female		Total		P value
	N=1077 (73.8%)		N= 382(26.2%)		N= 1459 (100%)		
	Frequency	Percent	Frequency	Percent	Frequency	Percent	
Gastrointestinal System disorders	466	43.3	244	63.8	710	48.8	<0.01
Genito-urinary system disorders	541	50.2	121	31.7	662	45.4	<0.01
CNS disorders	19	1.8	5	1.3	24	1.6	>0.05
Circulatory system disorders	21	1.9	5	1.3	26	1.8	>0.05
Musculoskeletal disorders	24	2.2	5	1.3	29	1.9	>0.05
Endocrine disorders	1	0.1	1	0.3	2	0.1	>0.05
Skin disorders	5	0.5	1	0.3	6	0.4	>0.05

Table 3: Different types of surgical procedures performed in the study cohort

Type of surgery	Number of patients
Cleft lip and palate repair	522
Herniotomy	451
VP shunting, removal and revision	24
Electrocauterization	10
Colostomy	39
Excision	86
Anal procedures	37
Conservative	6
Swenson Pull-Through Procedure	19
Orchiopexy	41
Others	224

Table 4: Complications during treatment of the anomalies

Type of complication	Number of patients
Death	3
Wound infection	1
Wound dehiscence	2
Recurrence after surgery	14
Bladder injury	1
Bad recovery	5
Failed treatment	5
Cerebrospinal Fluid (CSF) leak	1
Retention of urine	1

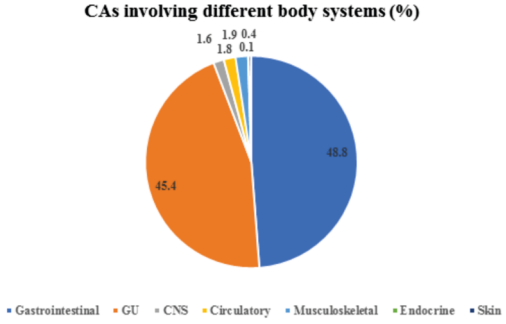

Figure 1: Pie chart showing congenital anomalies involving the different body systems

4. DISCUSSION:

Congenital anomalies are responsible for a staggering 57.7 million disability-adjusted life years (DALYs) lost worldwide.[11] Prevalence studies of congenital anomalies can help establish the baseline rates, document epidemiological changes over time, and identify any clues to the etiologies. Major congenital malformations have shown large variations in prevalence rates across worldwide, ranging from less than 100 to over 450 per 10,000 births.[12]

The present study showed that among children (0-14 years) reported with CAs over a period of twelve years, 73.9% were males and 26.10% in females. A similar higher prevalence in males was also found in another retrospective study conducted in El Shatby University Hospital.[1] In the current study, gastrointestinal and genitourinary system anomalies constituted 48.8% and 45.4% of the total CAs, respectively. Glassgow and Clyde surveillance study conducted in 2015–2016 recorded much lower gastrointestinal anomalies (9%) while musculoskeletal anomalies constituted 25%. Circulatory system anomalies constituted 1.8% of the total CAs in the current study. One surveillance study (Glasgow and Clyde) found that the prevalence of circulatory system anomalies was 13.9%, while another record (EUROCAT) showed comparatively higher figures for circulatory system anomalies, i.e.,35%. The percent of genitourinary anomalies was much higher than that of the EUROCAT records (45.4% of total CAs). [1]

Congenital anomalies are a significant cause of morbidity and mortality among infants and children under 5 years of age. Another study from North-Eastern India by Baruah *et al* has found that prevalence of congenital malformations was 1.2 % of the total live births. The distribution of malformation was predominant among males than in females (60.67 vs 37.37 %; p<0.05), similar to our present study. [6]

In our study almost half of the congenital anomalies are related to gastrointestinal anomalies, and the orofacial clefts have been considered under this category. Globally, the prevalence rate of Orofacial clefting (OFC) was suggested to be 1.5/1000 live births.[13]

Inguinal and scrotal swellings in children form the major part of the surgical conditions which require intervention. There were large number of congenital hernias observed in our cohort, next only to orofacial defects. Inguinal hernia repair is generally the most common operation performed in the pediatric age group. Various studies have reported an incidence of 3.5 to 5.0% for inguinal hernias in full-term infants and 44 to 55% in premature and Low Birth Weight (LBW) babies. [14,15]

Presently inguinal hernia is managed by herniotomy on a daycare basis. Laparoscopic hernia repair is generally conducted in adults, and there is little or no indication to use this technique in pediatric age group. There is a lack of pertinent data about the estimates of the surgical burden of disease [16] and more so about the pediatric surgical disease.[17] The burden of congenital anomalies falls very high in low-income countries (LICs) and low- and medium-income countries (LMICs), where 94 percent of anomalies occur.[18]

Higher fertility rates translate to higher birth rates and more children born with anomalies. Higher disease incidence is also attributed to higher micronutrient and macronutrient deficiencies, exposure to teratogens, prevalence of intrauterine infection, and self-medication with unsupervised drugs or traditional remedies. Some CAs can be easily repaired while others require staged, or multioperation, surgical interventions, and delays in treatment may result in lifelong illness, disability, and poor quality of life. There is paucity of surgical resources in LICs and LMICs. Anomalies resulting in visible deformity—such as clubfoot and cleft lip—also cause stigma, which can trigger abandonment or infanticide. Invisible deformities that result in chronic disability can lead to similar outcomes. Improving the pediatric surgical capabilities of LICs and LMICs will dramatically reduce this burden. Because children are the future economic engine powering the development of these countries, the value of investing in surgical care for children extends beyond Disability-adjusted life years((DALYs) averted to encompass the future socioeconomic well-being of LICs and LMICs themselves. It is critical to address the gaps in knowledge that impede the development of effective care systems.[19]

The world health organization (W.H.O.) and many other international bodies are also addressing the burden of congenital anomalies and their outcomes and are prioritizing this as a public health issue. The main focus is on the early recognition and development of new surgical techniques. Improvement in training facilities and monitory grants may also be beneficial for improving the outcome in developing countries.[3] There is a need for strong preventive measures for CAs. Measures that need to be taken to decrease the number of CAs in future include maternal care awareness during pregnancy, educational programs on CAs and the consequences of consanguineous marriages. There should be stress on improving nutritional status of women like general nutrition, ensuring adequate intake of specific micronutrients including folic acid, iodine, and iron; and removing harmful substances from the diet, especially alcohol, which may damage the developing embryo or fetus. Folic supplementation during the periconception period (three months before and after conception) can be useful. Studies suggest that 70% of neural tube defects can be prevented by the intake of daily dose of 400 g synthetic folic acid for women of childbearing age.[9]

The strengths of this study are long duration of enrollment and a large sample size.

Limitations

The major limitation of this study was its retrospective design. Moreover, we could not establish the risk factors of the CAs in this study due to the multiple sources of the data and lack of complete data availability across the study population to establish the relationship.

5. CONCLUSION:

The present study gives an overview of pattern of congenital anomalies in a town of Arunachal Pradesh over a long period of time. Surveillance and monitoring of congenital conditions are important for identifying patterns of malformations. A nationwide surveillance can recognize the disease burden in pre- and post-natal period and related risk factors. This will be helpful for strategic planning to improve the outcomes.

CONFLICTS OF INTEREST

The authors listed immediately below certify that they have no affiliations with or involvement in any organization or entity with any financial interest or nonfinancial interest in the subject matter or materials discussed in this manuscript.

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ETHICAL APPROVAL

The study obtained ethical clearance from the Institutional Ethics Committee, (Tomo Riba Institute of Health and Medical Sciences) TRIHMS, Naharlagun, Arunachal Pradesh.

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