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MORBIDITY AND MORTALITY PROFILE OF NEONATES WITH MAJOR CONGENITAL MALFORMATIONS IN A TERTIARY CARE HOSPITAL OF SOUTHERN RAJASTHAN

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ABSTRACT

Background- A malformation is a structural defect arising from a localized error in morphogenesis that results in the abnormal formation of a tissue or organ. Early intrauterine period (between 3rd to 8th weeks of gestation) is the vital period of life for the normal development of organs. Any insult within this period may result in congenital abnormalities. Congenital anomalies account for 8–15% of perinatal deaths and 4-8% of neonatal deaths in India.

Aims and Objectives-To study the morbidity and mortality profile of neonates born with major congenital anomalies in Southern Rajasthan.

Materials and Methods- The present study was a descriptive observational study conducted between January 2021 to December 2021 at NICU of teaching hospital of Southern Rajasthan including babies admitted for CMF during neonatal period.

Results- Incidence of Major CMF in our setting was 0.63% (86 of 13448 inborn live births). Out of 253 neonates admitted with CMF, most common anomalies were of G.I. system (60%) followed by CNS (21.7%) and multisystem involvement (7%). Tracheoesophageal fistula, (23.7%) Anorectal malformations (19.7%) and meningomyelocele (15.8%) constituted major chunk of the malformation. Out of 253 babies 71.5% were operated. Among neonates operated 51.5% babies developed septicemia,9%anaemia and 3% wound dehiscence postoperatively. Mean duration of hospital stay was 7 days with 40% babies being hospitalised for >10 days. Overall case fatality rate of CMF was 19.76%. Mortality rate of TEF and ARM was 18.6% and 15% respectively.

Conclusion-Mortality rate in CMF remains very high despite the availability of surgical intervention. Owing to its high incidence and high mortality Major congenital malformations take a heavy toll on health care system. Timely surgical intervention is of prime importance for the prognosis.

Keywords-Major congenital malformation (CMF), Tracheoesophageal fistula (TEF), Anorectal malformation (ARM).

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INTRODUCTION

Congenital anomalies can be defined as structural or functional defects that occur during intrauterine life and can be identified prenatally, at birth or sometimes detected in later infancy. Anomalies include malformations and dysplasia.[1]

A malformation is a structural defect arising from a localized error in morphogenesis that results in the abnormal formation of a tissue or organ. Dysplasia refers to the abnormal organization of cells into tissues.[2]

Early intrauterine period (between 3rd to 8th weeks of gestation) is the vital period of life for the normal development of organs. Any insult within this period may result in congenital abnormalities.[3]

Various factors have been identified for causation including monogenic/chromosomal aberrations, maternal infection, maternal illness, uterine environment, medication, nutritional and sporadic sequence.[4]

In earlier days it was thought that malformations were caused by monogenic defects in 7-8% of patients; chromosomal anomalies in 6%-8%; multigenic defects in 20%; and known environmental factors, such as maternal diseases, infections, and teratogens, in 6–7%. In the remaining 60–70% of patients, malformations were classified as caused by unknown etiologies.[2]

Prevalence of congenital anomalies vary from country to country due to the various racial, socio-cultural and ethnic influence. In the Middle East where consanguineous marriage is common, the prevalence of congenital anomalies is 2-2.5%. The overall incidence of congenital malformation in various studies range from 1-3%. [5]

Congenital anomalies account for 8–15% of perinatal deaths and 4-8% of neonatal deaths in India. The outcome of the disorder will depend on complex interactions between the prenatal deficit and the postnatal environment. Congenital anomalies can result in long-term disability, which may have significant impacts on individuals, families, health-care systems, and societies.[6]

This study was conducted to know the morbidity and mortality profile of babies with major congenital malformations.

MATERIALS AND METHODS

This was a Descriptive observational study conducted between January 2021 – December 2021 at NICU of Tertiary care teaching hospital of southern Rajasthan. After getting ethical committee clearance all the neonates admitted with major congenital anomalies admitted to NICU (Inborn and referred) were included in the study. Isolated minor anomalies (polydactyly, preterm PDA, etc.) were excluded from the study. Detailed maternal history including sociodemographic profile, antenatal visits, antenatal ultrasonography, Iron and folic acid supplementation, maternal history of drug addiction, smoking, alcohol intake and radiation exposure was collected. History of consanguinity, family history of malformations was asked. Detailed birth history including type of delivery, complications during delivery, need for resuscitation, APGAR score were noted. Anthropometry of the baby including birth weight, length and head circumference and other relevant information were recorded in a predesigned proforma,

Detailed general Physical examination including head to toe examination, vital parameters, evaluation of the baby for the anomalies was done. All orifices were examined carefully for patency by relevant means. Routine investigations, Imaging, genetic testing was done as and when necessary. Babies were stabilized according to the type of malformations as per NICU protocols. Newborns were assessed for operability by pediatric surgeon. All babies were followed up after surgery till discharge/death. Postoperative complications were managed accordingly.

RESULTS

During the study period total 13901 babies were delivered in our institute, out of which 759(5.46%) were still born, out of total 13448 live births, 86 babies had major congenital malformations. So, the incidence of major CMF was 0.63% in our institute.

After analyzing the data, it was found that out of 253 mothers, majority were in the age group of 20-24 years (47.8%) followed by, 25-29 years (35.37%). Majority of patients were from Rural background (94%) belonging to Upper lower class (43%) and lower Socioeconomic Status (40%). There were 53.7% primi women with 61.6% of pregnant women having visited ANC >4 times. In addition, only 2 cases (0.79%) were product of consanguineous marriage while 2 cases had first degree relatives with congenital anomalies. Out of all only 8.7% (22) anomalies were detected by antenatal USG.

Among 253 babies admitted for congenital anomalies,54% (136) were Male, 44.6% (112) were female,0.79% (2) were ambiguous. Majority of the cases were referred 68% (167) from periphery while 32% (86) babies were born in Dept of Obstetrics and Gynecology of same institute.

Looking system wise, majority of them had GI malformations 163 (64%), followed by CNS 58(22.7%), musculoskeletal 11(4.36%), genitourinary 8(3.1%), respiratory 7(2.7%), CVS 6(2.3%), and 18 babies had multisystem malformations. (Table 1)

Out of total 163 babies with GI anomalies 60 (37.4%) babies had Tracheoesophageal Fistula (TEF), 49(30%) had Anorectal malformations (ARM),15(9.2%) had Gastroschisis, 6(3.68%) had Omphalocele ,6(3.68%) had Jejunal atresia, 4(2.45%) had intestinal Obstruction, 4(2.45%) had cleft lip and palate, <2% babies had Hirschsprung, duodenal atresia, malrotation of gut, colonic atresia each.

Out of 58 babies with CNS anomalies, majority i.e., 40 babies had Meningomyelocele (68.9%) followed by encephalocele in 10(17.25%), Hydrocephalus in 5 babies (8.62%), Spina Bifida in 2(3.45%), Anencephaly in 1 baby (1.72%).

Among 253 cases Musculo-skeletal Anomalies were present in 11 cases. Out of which majority (45.45%) had Sacrococcygeal Teratoma, 27.27% had CTEV, 18.18% had B/L Upper Limb Deformity and 9.09% had genu recurvatum.

Amidst 253 cases Genitourinary System Malformation were present in 8 cases. Among them majority (5) had Recto Urethral Fistula, 2 had Ambiguous Genitalia, 1 had Exstrophy Bladder.

Out of 7 babies with respiratory system involvement 6 (85.5%) had Congenital Diaphragmatic Hernia,1 (14.5%) had Eventration of diaphragm.

Among 6 CVS malformations 3(50%) had Cyanotic congenital heart disease,2(33.3%) had Ventricular Septal Defect,1(16%) had complex congenital heart disease.
Out of 253 babies 180(71.5%) were operated, 65 (25.6%) were operable but could not be operated, 7(2.7%) babies were managed conservatively, and 1(0.45%) baby was inoperable (Table 2).

Among neonates operated 51.5% babies developed septicemia,9% babies developed anaemia and 3% developed wound dehiscence postoperatively (Table 3).

Out of all babies 81(32%) had a NICU stay of 0-5 days, 22.9% had 6-10 days, 20.5% had 11-15 days, and 24.1% babies had a stay >15 days (Table 4)

Out of total 71 TEF and TEF with ARM, 27 (36%) babies were operated and discharged, 18.6% babies were operated and expired, 34% babies could not be operated and 10% babies expired before surgery. Among babies with ARM (50) alone 78% babies were operated for colostomy and discharged, 15% had colostomy and expired, 4% could not be operated and 2% babies were non operable.

Out of 253 babies with CMF 170(67%) babies were discharged/DOR, 19% (50) babies expired, 11% (28) babies Left Against Medical Advice, and 2% (5) babies were referred to higher centre (Table 5)

Table 1

System	Number	Percentage
Gastrointestinal	163	64
CNS	58	22.7
Musculoskeletal	11	4.36
Genitourinary	8	3.1
Respiratory	7	2.7
CVS	6	2.3
Multisystem	18	7.1

Table-1 System wise distribution of major congenital malformations

Table -2

Treatment modalities	Number	Percentage
Operated	180	71.15
Operable but not operated	65	25.69
Conservative	7	2.77
Inoperable	1	0.40

Table-2 Treatment done

Table 3

Postoperative complications	Number	Percentage
Septicemia	34	51.52
Anemia	24	36.36
Shock	6	9.09
Wound dehiscence	2	3.03

Table-3 postoperative complications

Table 4

Duration (Days)	Number	Percentage
<5days	57	31.67
5-10 Days	51	28.33
>10days	72	40.00

Table- 4 Postoperative stay

Table 5

Outcome	Number	Percentage
Discharge/DOR	170	67.3
Death	50	19.76
LAMA	28	11
REFER	5	2

DISCUSSION

The present study "A STUDY OF MORBIDITY AND MORTALITY PROFILE AMONG NEONATES BORN WITH MAJOR CONGENITAL ANOMALIES ADMITTED IN NICU OF A TERTIARY CARE CENTRE IN SOUTHERN RAJASTHAN" was conducted in NICU of Tertiary care centre of Southern Rajasthan, from January 2021 to December 2021.

During the study period total 13901 babies delivered in our institute, out of which 759(5.46%) were still born out of total 13448 live births, 86(0.63%) babies had major congenital malformations. So, the incidence of major CMF was 0.63%. This could be explained by the fact that most of the deliveries in our hospital were booked cases and regular antenatal visits and recommended number of Ultrasonography were done in most of the cases. In a similar study by Kokate P et al[7] and Vyas R et al[8] incidence was found to be 0.9% and 1.23% respectively.

Out of all admissions40(16.2 %) were inborn babies and213(83.7%) babies were outborn, this is because of deficiency of specialist doctors at periphery and most of the babies born with CMF are referred to our centre. Out of all 137(54.4%) babies were male, 116(44.67 %) were female and 2(0.7%) ambiguous similar M: F proportion was found in studies by Verma Sehra et al[9](55:43), Sohier et al [10](54:46) and Farkhanda et al [11](61:38). Male preponderance is due to overall higher proportion of male child deliveries and affected female children are left untreated and not brought to referral centres. We observed that 94% affected babies were from rural and tribal background and more than 80% belonging to lower socioeconomic strata. This is due to extreme poverty, illiteracy, lack of knowledge about antenatal screening, poor antenatal care and unavailability of specialist doctors in these areas of southern Rajasthan.

Among of 253 babies with CMF 71.15% babies were operated, 25.09% babies were operable but could not be operated, 2.7% babies were managed conservatively and 0.4% babies were inoperable. Babies could not be operated were either brought in terminal stage, on mechanical ventilator or with multiple congenital anomalies or absconded due to long waiting list for OT.

(Table 2) 90 babies (51.5%) out of total 180 operated babies developed sepsis, 9% (16) babies each developed anaemia and shock and 3% (5) babies had wound dehiscence. High

incidence of sepsis was due to high patient load and long waiting leading to delayed intervention, majority of the babies being referred cases, higher incidence of sepsis in out born babies. Anaemia was due to blood loss during surgery.

(Table 3) Out of all babies 81(32%) had a NICU stay of 0-5 days, 22.9% had 6-10 days, 20.5% had 11-15 days, and 24.1% babies had a stay >15 days (Table 4). Prolonged duration of hospital stay was due to complications associated with the surgery and delayed post operative healing. Prolonged hospital stay was associated with increased mortality.

Out of all babies 67.3% were discharged successfully, 19.7% babies died, 11% babies Left against Medical Advice (LAMA), and 2% babies were referred (Table 5). The referred babies were case of exstrophy bladder (1) and Critical cyanotic CHD (4) due to unavailability of Paediatric Cardiothoracic surgeon and Paediatric urologist in our hospital. In a similar study by Ritu Vyas et al[8], 50% babies were discharged, 34% babies expired, 7% babies were referred and 9% babies DAMA. In a study by Tenali et al[12], out of 40 babies born with CMF 5 babies expired soon after birth, 3 babies with NTD were operated with intact development at follow up, 4 babies with cardiac anomalies were operated out of which 3 survived. Our study in concordance with other study showed high rate of mortality among anomalous babies in comparison to non-anomalous babies due to life threatening nature of the anomalies, delay in intervention due to high patient to surgeon ratio, high incidence of postoperative complications. Most common cause of mortality was sepsis among operated babies.

A special mention of GI anomalies commonly admitted in the nursery, out of all TEF and TEF with ARM babies 36% babies were operated and discharged, 34.6% babies could not be operated, 18.6% babies were operated and expired, 10.65 %babies were expired before surgery.

Out of all ARM babies 78.27% babies were operated for colostomy and discharged, 15.2% babies had colostomy and expired, 4.34% babies could not be operated and 2.17% babies were inoperable. ARM being one of the most common CMF if identified early and operated has good chances of survival.

CONCLUSION

Congenital anomalies make an important contribution to neonatal morbidity and mortality. They remain a leading cause of neonatal death all over the world. Highest number of neonates born with congenital anomalies were due to gastrointestinal defects, followed by CNS defects and multisystem defects. Survival of affected babies can be significantly improved if corrective surgery done at right time. High mortality is due to delay in identification, lack of specialist doctors at periphery, awareness about surgical mode of treatment among general population and postoperative complications.

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REFERENCE

1. World Health Organization. Congenital Anomalies Leading causes of neonatal death. Available from: http://www.who.int/news-room/ fact-sheets/detail/congenital-anomalies. WHO; 2000-2016.

2. Anne M. Slavotinek Dysmorphology; NELSON TEXTBOOK OF PEDIATRICS, 21st edition, Elsevier Publishers2020;1:986-89.
3. Ravikumar M, Bhat V, Oumachigui A, Perinatal mortality trend sin referral hospital," Indian J Pediatr, 1996;63:357–361,.
4. Behrman RE, Kliegman RM, Dysmorphology; NELSON TEXTBOOK OF PEDIATRICS, 21st edition, Elsevier Publishers2020;1:988.
5. Mashuda F, Zuechner A, Chalya PL, Kidenya BR, Manyama M. Pattern and factors associated with congenital anomalies among young infants admitted at the Bugando Medical Centre, Mwanza, Tanzania. BMC Res Notes. 2014; 7:195.
6. Taksande A, Vilhekar K, Chaturvedi P, Jain M. "Congenital malformations at birth in Central India: A rural medical college hospital based data," Indian J Hum Genet. 2010; 16:159–163.
7. Kokate P, Bang R. Study of congenital malformation in tertiary care centre, Mumbai, Maharashtra, Int J Reprod Contracept Obstet Gynecol. 2017;6:89-93.
8. Vyas R, Verma S, Malu VK. Distribution of congenital malformations at birth in a tertiary care hospital in North-Western Rajasthan. Int J Reprod Contracept Obstet Gynecol 2016;5:4281-4
9. Verma SV, Sehra RN. A Study on Incidence of Various Systemic Congenital Malformations at Birth in a Tertiary Care Hospital of Northwestern Rajasthan. J South Asian Feder Obst Gynae; 2016; 8:309-313.
10. AbouEl-Ella SS, Tawfik MA, Abo El-Fotoh WMM, Elbadawi MA. Study of congenital malformations in infants and children in Menoufia governorate, Egypt. Egyptian J Med. Hum Gene. 2018 Oct;19(4):359–65.
11. Farkhanda T, Sabir S, Gul H. Prevalence and Distribution of Congenital Malformations in Newborns in Tertiary care Hospital. Ann. Pak. Inst. Med. Sci. 2017; 13:242-245.
12. Tenali ASL, Kamlakannan S, Jayaraman K. Spectrum of congenital anomalies of neonates in a tertiary care hospital in Southern India. Int J Contemptr Pediatr. 2018; 5:314-9.
13. Egbe A, Uppu S, Lee S, Stroustrup A, Ho D, Srivastava S. Congenital malformations in the newborn population: A population study and analysis of the effect of sex and prematurity. Pediatr Neonatol 2015; 56:25–30.
14. Parmer A, Rathod SP, Patel SV, Patel SM. A study of congenital anomalies in newborn. NJIRM 2010; 1:13–7.
15. Khatemi F, Mamoori GA. Survey of congenital major malformations in 10/000 newborns. Iran J Pediatr 2005; 15:315–20.

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