Prevalence of neurological malformation in newborns at a tertiary care center in Rajasthan, India

Jaiprakash Narayan[1], Suchitra Narayan[2], Anil Kumar Jain[3], Pukhraj Garg[3], Bhag Singh Karnawat[3]

From [1]Associate Professor, [3]Senior Professor, Department of Pediatrics, [2]Assistant Professor Department of Obstetrics and Gynecology, Jawaharlal Nehru Medical College, Ajmer, Rajasthan, India

Correspondence to: Dr. Suchitra Narayan, 25 J. C. Nagar, Makarwali Road Vaishali Nagar Ajmer - 305 001, Rajasthan, India. E-mail: narayan_jaiprakash@yahoo.co.in

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ABSTRACT

Introduction: Many newborns die every year due to various congenital anomalies and those who survive, suffer from long-term morbidity. To deal with birth defects, a large proportion of health resources and workforce is required. Among the various congenital anomalies, neurological birth defects are the leading type. **Objective:** The objective of the study was to document the epidemiological features and prevalence of congenital neurological anomalies in rural areas. **Materials and Methods:** This retrospective study was conducted in the department of pediatric of a medical college of Rajasthan. The study population included live births born in our institution between 1990 and 2018. A register was maintained after delivery to document the particulars of neonates with a congenital birth defect and their mothers. We had taken neonatal record from this register and analyzed the data of the past 28 years, retrospectively. **Results:** Of 241,848 live births, 6623 cases were identified with a primary diagnosis of one or more congenital anomalies giving a prevalence rate of 274/10,000 live births (95% confidence interval [CI]: 262.2–285.8). Anomalies of the nervous system were the second common defects, accounting for 19.95% of the birth defects just after the heart disease anomalies (21.65%). Neurological anomalies were diagnosed in 1321 neonates. The prevalence of neurological anomalies was 54.62/10,000 live births (95% CI: 49.75–59.49). The most common neurological anomaly was myelomeningocele/meningocele (spina bifida cystic) found in 64.87% of cases followed by Chiari malformation (7.72%), encephalocele (6.89%), microcephaly (6.88%), hydrocephalous (1.43%), and spina bifida occulta (1.43%). **Conclusion:** Neurological anomalies were among common congenital anomalies and considerable cause of mortality and morbidity. Myelomeningocele/meningocele (spina bifida cystic) was the most common neurological anomaly.

Key words: *Congenital anomalies, Myelomeningocele, Neurological anomalies*

A congenital malformation (CM) or birth defect is defined as a structural or chromosomal malformation with a significant impact on the health and development of a child [1]. With regard to morbidity, CMs account for 12% of all pediatric hospitalizations [2]. This subset of patients with CMs has longer hospital stays and incurs higher hospitalization costs, compared to other patients [3-5]. Studies published worldwide report a birth prevalence of CM that ranges 20–55/1000 live births with significant variation, depending on the demographics of the study population, the study design, and the method of case ascertainment [1,4,6-9]. Approximately 21% of these anomalies involve the central nervous system (CNS), constitute one of the most common birth defects, second only to heart defects [10].

Literature shows that neural tube defects (NTDs) are among the most common congenital abnormalities, but the prevalence varies between countries and races [10]. In the UK, anencephaly and spina bifida are of approximately equal prevalence and together make up 95% of all NTDs [11]. Congenital anomalies account for 8–15% of perinatal deaths and 13–16% of neonatal deaths in India [12,13]. Various Indian studies reported the prevalence of congenital anomalies from 160 to 430/10,000 live births [14-21]. We planned this retrospective cross-sectional study to know the prevalence of CM, incidence, and prevalence, distribution of neurological malformation at tertiary care center at central Rajasthan, India.

MATERIALS AND METHODS

This retrospective cross-sectional study was conducted in the Department of Pediatrics of a tertiary care teaching institution of Rajasthan, India. The study population included live births born in our institution between 1990 and 2018. This hospital serves both urban and rural population and is a tertiary referral hospital of central Rajasthan. The study was planned and permission was taken from the Institutional Ethical Committee and Institutional head to use hospital data for research.

A register (congenital anomalies register) was maintained by the resident doctors to document particular of neonates with a

congenital birth defect and their mothers after delivery. In this register, resident doctors filled particulars of mothers and their babies, risk factors if available, weight and gestational age of mother, history of consanguinity, referral and outcome of baby, and duration of stay.

Authors had taken the neonatal record from this register, filled all the data in a Microsoft Excel sheet regarding gestational age, birth weight, parity, history of consanguinity, sex of the child, and other relevant histories, the various congenital anomalies were also noted down. Authors had taken total live birth data from the register maintained by labor room resident doctors. Finally, the data of neonates with CMs of the past 28 years were analyzed.

RESULTS

Of 241,848 live births, 6623 cases were identified with a primary diagnosis of one or more congenital anomalies; out of them, 54.91% were male and 44.83% of female. It gives a prevalence rate of 274/10,000 live births (95% confidence interval [CI]: 262.2–285.8). Anomalies of the nervous system were the second common defects, accounting for 19.95% of the birth defects just after the heart disease anomalies (21.65%).

Neurological anomalies were diagnosed in 1321 neonates; of these, 53.7% were male and 46.18% female (Table 1). The prevalence of neurological anomalies was 54.62/10,000 live births (95% CI: 49.75–59.49). Among live-born children having congenital neurological anomalies, 62.9% of male and 63.77% of female children were born vaginally.

As shown in Table 2, the neurological anomalies were more common in <2.5 kg children and also more in male as compared to a female child, whereas in >4 kg weight female children were more affected than male children.

Table 3 is indicating that the neurological anomalies are more common in preterm live birth. About 49.67% of neurological anomalies were found in neonates of gestational age between 37 and ≤42 weeks in female, while in the similar age bracket, the neurological anomalies in male were 47.53%.

Outcome of these babies is presented in Table 4 which shows that around 4% liveborn died during a hospital stay due to congenital neurological anomalies. In general, children with critical congenital anomalies or if parents are not willing for the further treatment they choose the option of LAMA and these children would die on their way to home or at home. If these statistics are included and these children are taken as expired, the children mortality figure will see an increase of 8.4%. The mortality/survival data were not available of those children who were referred to the higher center or other departments for further management to know about the occurrence of their mortality rate during the surgery or post-operative period.

The most common neurological anomaly was myelomeningocele/meningocele (spina bifida cystic) found in 64.87% of cases followed by Chiari malformation (7.72%), encephalocele (6.89%), microcephaly (6.88%), hydrocephalus (1.43%), and spina bifida occulta (1.43%). The prevalence of congenital anomalies year wise was varying in between 2.2 and 3.75% as presented in Table 5.

Table 1: Distribution of children according to gender

Gender	Total anomalies (%)	Neurological anomalies (%)
Male	3637 (54.91)	709 (53.7)
Female	2969 (44.83)	610 (46.18)
Ambiguous genitalia	17 (0.0026)	2 (0.0015)
Total	6623	1321

Table 2: Distribution of children according to birth weight

Weight birth (kg)	Male (%)		Female (%)		Ambiguous genitalia (%)	
	Total anomalies	Neurological anomalies	Total anomalies	Neurological anomalies	Total anomalies	Neurological anomalies
<1	281 (7.726)	69 (9.73)	237 (7.98)	54 (8.85)	4 (23.53)	1 (50)
1–1.449	292 (8.02)	98 (13.8)	209 (7.03)	67 (10.98)	3 (17.65)	1 (50)
1.5–2.449	1309 (35.99)	289 (40.76)	1096 (36.91)	221 (36.23)	6 (35.29)	0
>2.5	1698 (46.68)	302 (42.59)	1337 (45.03)	251 (41.15)	3 (17.65)	0
>4	57 (0.0156)	13 (0.018)	90 (3.03)	17 (2.79)	1 (5.88)	0
Total	3637	709	2969	610	17	2

Table 3: Distribution of children according to gestational age

Gestational age (weeks)	Male (%)		Female (%)		Ambiguous genitalia (%)	
	Total anomalies	Neurological anomalies	Total anomalies	Neurological anomalies	Total anomalies	Neurological anomalies
<32	371 (10.2)	82 (11.56)	355 (11.96)	62 (10.16)	6 (35.290)	1 (50)
32–≤37	1192 (32.77)	269 (37.94)	419 (14.11)	227 (37.21)	4 (23.53)	1 (50)
37–≤42	1871 (51.44)	337 (47.53)	1705 (57.43)	303 (49.67)	7 (41.18)	0
42 and more	203 (5.58)	21 (2.96)	490 (16.50)	18 (2.96)	0	0
Total	3637	709	2969	610	17	2

Table 4: Distribution of children having neurological anomalies according to the outcome

Outcome	Male (%)	Female (%)
Discharged	489 (68.97)	391 (64.1)
Expired	28 (3.94)	23 (3.77)
LAMA	13 (1.83)	45 (7.44)
Referred	179 (25.25)	151 (24.75)
Total	709	610

Table 5: Types and percentage of congenital neurological anomalies

Congenital anomaly	Number of patients (%)
Anencephaly	23 (1.74)
Encephalocele	91 (6.89)
Myelomeningocele/meningocele (spina bifida cystic)	857 (64.87)
Chiari malformation	102 (7.72)
Diastematomyelia	5 (0.37)
Holoprosencephaly	11 (0.83)
Dandy–Walker syndrome	27 (2.084)
Microcephaly	91 (6.88)
Megalencephaly	6 (0.45)
Porencephaly	9 (0.68)
Hydranencephaly	9 (0.68)
Schizencephaly	7 (0.52)
Lissencephaly	2 (0.15)
Hypoplasia/aplasia of the corpus callosum	4 (0.3)
Septo-optic dysplasia	3 (0.23)
Aqueduct stenosis	7 (0.52)
Hydrocephalous	19 (1.43)
Arachnoid cyst	8 (0.6)
Spina bifida occulta	19 (1.43)
Syringomyelia	7 (0.52)
Heterotopias	5 (0.37)
Polymicrogyria	4 (0.3)
Agenesis of the corpus callosum	3 (0.23)
Hypoplasia of vermis	2 (0.15)

DISCUSSION

In the present study, 6623 cases were diagnosed with congenital anomalies; out of them, 54.91% were male and 44.83% of female. Egbe *et al.* also reported CM in 51% of male similar to the present study [2]. While in neurological anomalies, 53.7% were male and 46.18% female. Neurological anomalies were more common in <2.5 kg children and also more in male as compared to female children while in >4 kg weight female children were more affected than the male children. Neurological anomalies are more common in a preterm live birth in the present study similar to other studies [2]. Among live birth having congenital neurological anomalies, the vaginal method of delivery was observed in 62.9% of male and 63.77% of female. Around 4% liveborn die regularly due to the congenital neurological anomalies and 25% are referred to pediatric surgery.

The most common neurological anomaly was myelomeningocele/meningocele (spina bifida cystic) followed by Chiari malformation. The prevalence of congenital anomalies was in between 2.5 and 3.3%. Of 241,848 live births, 6623 cases had congenital anomalies with a prevalence rate of 274/10,000 live births. Among hospital studies, which included data on both live births and stillbirths, anomalies of the CNS were most frequently reported followed by anomalies of the musculoskeletal system (75.85/10,000 births [95% CI 58.80–92.90] and 65.64/10,000 births [95% CI 52.97–78.31], respectively). Among live births, anomalies of the musculoskeletal system were highest in both hospital (79.38/10,000 live births [95% CI 32.32–126.44]) and community settings (65.88/10,000 live births [95% CI 23.13–108.63]). The corresponding prevalence of CNS defects was lower (28.93/10,000 live births [95% CI 13.64–44.22] for hospital-based studies and 26.19/10,000 live births [95% CI 15.55–36.83] for community-based studies) [22].

The global incidence of malformations was 3.67%, according to other reports in literature that describes an incidence of 2–5% of significant structural anomalies similar to the present study [23]. Anomalies of the nervous system were the second common defects, accounting for 19.95% of birth defects just after the heart diseases anomalies (21.65%) similar to the other studies [1]. In accordance to the present study, neurological and CM were more common in preterm and large for gestational age [24].

In some studies, CNS malformations are described as the most common type of malformation, comprising up to 13% of cases, second only to congenital heart disease [1]. Total neurological anomalies were 1321. The prevalence of neurological anomalies was 54.62108/10,000 live births (standard deviation – 17.03, 95% CI: 49.75–59.49).

CONCLUSION

Neurological anomalies constitute significant percent of total CM and cause of neonatal mortality and morbidity.

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