ORIGINAL ARTICLE

Pattern of Congenital Malformations and their Neonatal Outcome at Sheikh Zayed Medical College/Hospital Rahim Yar Khan

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ABSTRACT

Objective: To highlight the pattern of congenital malformations in neonates admitted in a tertiary care Nursery and to evaluate their outcome after treatment.
Study design: Descriptive study.
Place & Duration of study: Department of Pediatrics Medicine & Surgery, Sheikh Zayed Medical College & Hospital, Rahim Yar Khan from January to December 2010.
Methodology: A total of 4270 patients were admitted in nursery. Neonates with congenital malformations were diagnosed by clinical examination and by appropriate investigations. Surgical intervention was done accordingly.
Results: Out of 4270 patients admitted in Nursery in one year, 645 patients (15%) were diagnosed as having congenital malformations. 335 were male and 302 were female & 8 were having ambiguous genitalia. 45 neonates died pre-operatively, 345 patients had surgery out of which 42 patients (12.1%) died in neonatal period, 217 were advised surgery after neonatal Period, and 38 patients refused surgery & were managed conservatively.
Conclusion: By improvement in antenatal, postnatal diagnosis, urgent referral to tertiary care center, early surgical intervention and intensive post operative neonatal care, most infants can be saved.
Key words: Congenital, Malformation, Neonate.

INTRODUCTION

Congenital malformations are morphologic defects that occur in the pre-natal period as a result of genetic mutations, chromosomal abnormalities and adverse intrauterine environment. These are present at birth & clinically manifest at any time in life. There is a wide variety of fetal abnormalities ranging from relatively minor abnormalities to major defects[1]. Minor anomaly usually involves non vital organs with or without any functional defects. Mostly they do not cause any distress in the newborn and usually do not need early correction especially in the neonatal period. As compared, major anomalies involve vital organs causing functional defects and are of significant cosmetic value, even life threatening. So they need immediate / early correction, if not corrected so, these anomalies could impair child's development.

Maternal ultrasonography can diagnose these anomalies prenatally in 2nd trimester of life [2,3] & intervention of certain congenital malformations like hydrocephalus, PUV, cleft lip, hydronephrosis in intrauterine life (FETENDO) are gaining popularity now a days. Neonatal surgical intervention is done usually soon after birth, these procedures are not only to restore the structure but also the function. Cosmetic effect is also improved.

The world wide incidence of congenital anomalies is estimated as 3-7%, but actual incidence varies widely between countries[4]. Many studies have been done in different parts of world as well as in Pakistan but no such study have been conducted in this area of Punjab. Sheikh Zayed hospital caters almost whole of southern Punjab & areas of Sind and Baluchistan province. The peoples over here belong to different cultures and have exposure during antenatal period to various environmental factors like medications as desire for a particular sex. So this study was done to document the pattern of congenital anomalies in neonates.

MATERIALS & METHODS

This descriptive study was carried out at Sheikh Zayed Medical College / Hospital, Rahim Yar Khan on all the neonates admitted in Nursery from January 2010 to December 2010. All cases with both major & minor congenital anomalies were enrolled. A thorough physical examination was performed &

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various imaging modalities i.e. Radiography, USG and CT scan were done where ever needed. The anomalies diagnosed on Pre-natal ultrasonography were confirmed clinically and by appropriate radio-diagnostic methods soon after birth. The neonates were managed accordingly. Post operative outcome in terms of morbidity & mortality was noted.

RESULTS

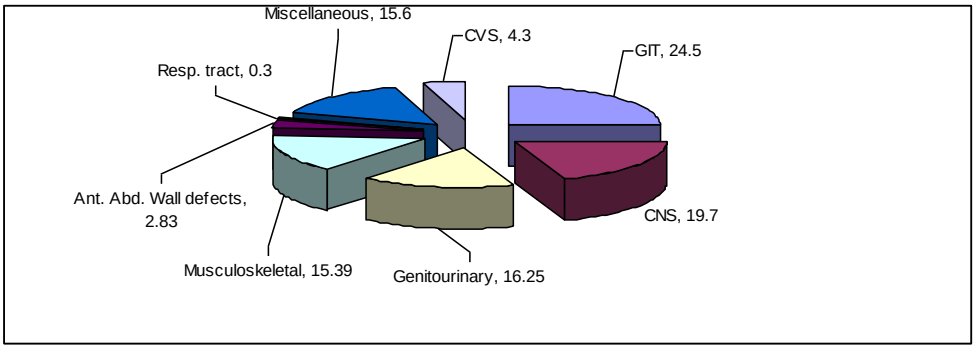
The total 4270 neonates were admitted during study period, out of which 645 patients had congenital malformations making prevalence as 15%. 478 patients had single anomaly and 167 patients had more than one anomaly, thus a total of 812 anomalies among 645 patients. 335 patients (51.9%) were male and 303 (46.9%) were female and 08 patients (1.2%) were having ambiguous sex. All patients were actively managed both medically and surgically. Reports of fetal ultrasonography were available in only 104 patients. Out of these, the anomalies were identified in only 68 patients while it was missed in 30 patients and incorrectly diagnosed in 06 patients. Based on clinical examinations and relevant investigations, all cases were categorized into organ specific involvement. The pattern of congenital anomalies in terms of frequency is shown in Table-I. The most common anomalies were of gastrointestinal tract, followed by CNS. Regarding outcome, 45 neonates died pre-operatively, 345 patients had surgery out of which 42 patients (12.1%) died in neonatal period, 217 were advised surgery after neonatal Period, and 38 patients refused surgery & were managed conservatively.

Table I: Results of the study

Type of congenital anomaly	No. of patients	%age
Gastrointestinal:		
Anorectal malformation	30	3.69
Intestinal atresia/stenosis	34	4.18
Jejunal	15	1.84
Illeal	13	1.60
Duodenal	06	0.7
H.D.	15	1.8
Meconium illeus	14	1.7
Malrotation	12	1.4
Annular Pancreas	03	0.3
Diaphragmatic hernia	06	0.7
Mesenteric cyst	06	0.7
TOF (Tracheo esophageal fistula)	17	2.0
Patent vitello intestinal duct	04	0.4
Cleft lip & palate	58	7.1
Lip	37	4.5
Palate	21	2.5
	Total = 199	24.50
CNS		
Meningomyelocele / Meningocele	73	8.9
Hydrocephalus	44	5.4
Encephlocele / En-encephaly	18	2.2
Sacrococcegeal teratoma	07	0.8
Occipital bursa	12	1.4
Frontocele	06	0.7
	Total = 160	19.70

Genitourinary:		
Hypospadias	28	3.4
Ambigous Genitalia	08	0.9
Inguinal hernia	29	3.5
Bilateral	10	1.2
Unilateral	19	2.3
Prune Belly Syndrome	04	0.4
PUV	14	1.7
Hydronephrosis	12	1.4
Ectopia vesicae	03	0.3
Patent urachus	03	0.3
Epispadias	06	0.7
Undescended testes	25	3.0
	Total =132	16.25
Musculoskeletal		
Club foot	68	8.3
Syndactly	28	3.4
Polydactyly	14	1.7
DDH	12	1.4
Craniosynostosis	03	0.3
Absence of radius	08	0.9
	Total =125	15.39
Anterior Abdominal Wall Defects:		
Omphalocele	17	2.0
Gastroschiasis	06	0.7
	Total =23	2.83
Respiratory tract		
Choanal atresia	03	0.3
Miscellaneous:		
Cystic hygromas	25	3.0
Torticollis	11	1.3
Haemangiomas	73	8.9
Ear deformities	10	1.2
Upper eyelid defect	08	0.9
	Total = 127	15.6
Cardiovascular system		
CHD (VSD, ASD, TOF, TGA)	35	4.3

Table II:



DISCUSSION

Our study indicates that congenital anomalies are important Pediatric problem, constituting 15% of total admissions in a neonatal tertiary care center. The high incidence of congenital anomalies at Sheikh Zayed Hospital Rahim Yar Khan is due to the fact that it caters almost whole of southern Punjab & areas of Sind and Baluchistan province, over here the incidence of congenital anomalies is more as compared to rest of the country and the world wide incidence[11].

The pattern of malformations is different in different regions of the world. In our study, the most

common pattern of anomalies was GIT defects. In three different studies from Iran, Musculoskeletal anomalies rank as the first [5,6,7]. While two studies from India also report the same [8,9]. Another Indian study reports CNS anomalies to be the most frequent [10]. All these studies are hospital based which may not reflect the overall status of problem. To get a better picture of problem, community studies should be under taken.

Although in our study, GIT anomalies are most common, but CNS anomalies also constitute a significant number of patients comparable to another study done before in this unit [11]. It also shows poor socioeconomic status and unawareness of peoples of this area about this problem.

A study performed in a Nigerian Teaching Hospital from 1991-2000 shows that gastro-intestinal anomalies were among the commonest type (45%) of congenital malformations followed by CNS anomalies (24.2%) favoring our results[12].

Prenatal ultrasonography leads to early detection of malformations and facilitates early surgical intervention. However fetal ultrasonography may not pick up all cases [13,14].

In our study, only 104 patients had got fetal ultrasonography and in only 68 patients where anomalies were identified that shows lack of awareness among peoples of this region and lack of competency among sonologists.

Emotional stress for parents is another important issue that needs counseling for early surgical intervention for better outcome. Another issue is termination especially for conditions incompatible with life that needs appropriate law. In our study, the main factor in post-operative mobility and mortality was delayed referral from peripheral centers because most deaths occurred in neonates referred from outside. The other factors were prematurity and infections. According to WHO, congenital anomalies are a major burden on underdeveloped countries and they are more concerned about this issue[15].

CONCLUSION

This study definitely helps to know the pattern of congenital anomalies and their neonatal outcome in this area. A nation wide surveillance can recognize the disease burden in pre & post-natal period and related risk factors to plan future strategies for prevention, early diagnosis and timely management.

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