The Incidence of Correctable Congenital Anomalies in Bahrain

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ABSTRACT

This prospective study was performed to examine the possible incidence of correctable congenital anomalies in Bahrain. The overall incidence of correctable surgical anomalies is 3.1% of live births excluding congenital heart diseases and congenital orthopaedic abnormalities. The most common urogenital anomalies are undescended testes with an incidence of 158 / 10,000 live births and hypospadias with an incidence of 15.3 / 10,000 live births. The most common gastrointestinal anomalies are gastrointestinal atresia with an incidence of 10.6 / 10,000 and Hirschsprung's disease of 2.9 / 10,000 live births. Anorectal malformation is very common, but biliary tract anomalies are extremely rare. Congenital inguinal hernia is very common with an incidence of 89 / 10,000 live births, but exomphalos, gastroschisis and exstrophic anomalies are also rare. The most common neurological anomalies are hydrocephalus and myelomeningocele with an incidence of 12.3 / 10,000.Cleft lip and palate are the commonest facial anomalies with an incidence of 11.5 / 10,000.

Congenital anomalies are important causes of neonatal mortality and morbidity. Knowing the extent of the problem will help us to plan our future health service in medical and surgical neonatology. The purpose of this communication is to present a prospective study of the incidence of correctable surgical anomalies in Bahrain, which might reflect the incidence in the Arabian Gulf region.

METHODS

Bahrain is an archipelago of islands with an area of 680 km^2 and a population of over 400,000. The annual birth rate is over 12,000 and 98% are hospital deliveries. The male to female delivery ratio is almost 1:1. Each newborn is routinely examined by a paediatrician within 24 hours after hospital delivery. The major congenital anomalies are referred to our Paediatric Surgical Unit at Salmaniya Medical Center, which is the only paediatric surgical unit on the island. We collected all major congenital anomalies treated in our unit from April 1980 to December 1988. There were almost 105,000 live births within this period. From the available data we studied the incidence of various congenital anomalies on the island. To know the incidence of undescended testes at birth we calculated all the undescended testes we operated upon and multiplied them by three, presuming from previous study[1] that two third of undescended testis will descend within one year of age. The incidence of hypospadias in this report excludes glandular hypospadias which we do not repair.

RESULTS

The overall incidence of correctable congenital anomalies excluding congenital heart disease and congenital skeletal deformities were 3.1% (Table I). The commonest congenital abnormalities were the urogenital anomalies. The most common urogenital anomalies were undescended testes, with an estimated incidence of 158 / 10,000 live births. The incidence of hypospadias (excluding glandular type) was 15.3 / 10,000, congenital hydronephrosis with pelviureteric junction obstruction 4.9 / 10,000 and congenital urethral obstruction 3 / 10,000 including posterior urethral valve 2.3 / 10,000 live births (Table II)

Gastrointestinal anomalies are the second most common group of congenital anomalies (Table III). The incidence of neonatal intestinal obstruction was 14 / 10,000, and Hirschsprung's disease 2.9 / 10,000 live births. The overall incidence of gastrointestinal atresia is 10.7 / 10,000, which includes oesophageal

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TABLE I

Total Incidence of Correctable Congenital Anomalies Excluding Congenital Heart Disease and Orthopaedic Anomalies

Anomaly	*incidence / 10,000*
Undescended testes	158
Hypospadias	15.3
Congenital hydronephrosis	4.9
Cong. urethral obstruction	3
Neonatal intestinal obst.	14
Congenital inguinal hernia	89
Diaphragmatic hernia	2.3
Exomphalos & Gastroschisis	1
Myelomeningocele & hydroceph.	12.3
Cleft lip & palate	11.5
Cong. lobar emphysema	1
Total	**312.3**

TABLE II

Types of Urogenital Anomalies between April 1980 to December 1988

Anomaly	*Number*	
Undescended testes	1,656	552.3
Hypospadias	153	
Congenital urethral obst.	31	
Congenital hydronephrosis PUJ obstruction	51	

TABLE III

Types of Gastrointestinal Anomalies between April 1980 to December 1988

Anomaly	*Number*
Neonatal intest. obst.	147
Hirschsprung's disease	30
Gastrointestinal atresia	112

atresia 2.6 / 10,000, duodenal atresia 1.8 / 10,000, small bowel atresia 1.4 / 10,000, and imperforate anus 4.5 / 10,000 live births. Colonic atresia is very rare, we have seen only one case within this period. Hepatobiliary anomalies were also rare, we have seen one biliary atresia, one choledochal cyst, and two gall bladder agenesis without biliary atresia as incidental laparotomy finding (Table IV).

Inguinal hernia is a common abnormality with an incidence of 89 / 10,000 live births. Abdominal wall defects were rare: congenital diaphragmatic hernia 2.3 / 10,000; exomphalos and gastroschisis 1 / 10,000. Exstrophy anomalies were also rare, we have seen three cases of cloacal exstrophy, three cases of cloacal anomalies two cases of bladder exstrophy, and two cases of epispadias.

The most common neurological abnormalities were myelomeningocele and hydrocephalus with an incidence of 12.3 / 10,000. Cleft lip and palate were the commonest craniofacial anomalies with an incidence of 11.5 / 10,000 live births. Congenital lobar emphysema was rare with an incidence of 1 / 10,000. Within this period we have seen seven cases of congenital adrenal hyperplasia and three cases of testicular feminization syndrome (Table V).

TABLE IV

Total Number of Gastrointestinal Atresia between April 1980 and December 1988

Type	*Number*
Oesophageal atresia	26
Duodenal atresia	19
Small bowel atresia	15
Colonic atresia	1
Anorectal atresia	47
Biliary atresia	1
Choledochal cyst	1
Gall bladder agenesis	2
Total	**112**

TABLE V

Total Number of Other Congenital anomalies between April 1980 to December 1988*

Anomaly	*Number*
Congenital inguinal hernia	934
Congenital diaphrag. hernia	24
Exomphalos and gastroschisis	11
Bladder exstrophy	2
Epispadias	2
Cloacal exstrophy	3
Cloacal anomalies	3
Myelomeningocele and hydrocephalus	129
Cleft lip and palate	121
Congenital lobar emphysema	10
Congenital adrenal hyperplasia	7
Test. feminization syndrome	3

* Total number of correctable congenital anomalies seen is 3287

DISCUSSION

The over-all incidence of congenital anomalies is almost constant throughout the world[2], but the incidence of each anomaly varies very considerably between one place and another. For example, anencephaly and spina bifida appear to be much less common in the African and Asian than amongst European[3]. In Japan, the incidence of cleft lip and palate is reported to be 21/10,000[4], more frequently than amongst western communities. Regional differences are remarkable in many countries. The reported incidence of neural tube defects is ten times more in Bombay than Calcutta[2]. In Great Britain the incidence of neural tube defects varies between 72/10,000 in Ireland, 56/10,000 in Scotland[5] and 36/10,000 England[6].

For practical purposes two quite different figures of incidence may be needed. To ascertain all malformed infants within a community and to determine from this the overall incidence is important for epidemiologist to know the extent of the problem. But for organizing neonatal medical and surgical service, the relevant figures are the live-born infants with treatable deformities. For example, in planning neonatal surgical services for children with myelomeningocele the total incidence would be misleading, for as many as one-quarter of these may be still-born[6]. Again, a figure for total incidence of congenital diaphragmatic hernia would be valueless to the clinician. More than half of these anomalies occur in anencephalics or infants with anomalies incompatible with survival[1]. There is no doubt knowing the number of all malformed infants will allow the epidemiologist to understand the magnitude of the problem in the community and the amount of efforts needed to prevent it. It will direct our research to the etiology and possible early treatment.

It is generally accepted[2] that to study the incidence of congenital anomalies, a base of at least 50,000 live births is required to know the extent of problem. In our attempt to study the incidence of congenital anomalies in Bahrain, we reviewed all the correctable congenital anomalies which were treated at our paediatric surgical unit in the past nine years. The number of these anomalies might refelct the incidence of the correctable congenital anomalies in Bahrain, because of: the size of the island; the 98% hospital delivery; the easy referral; and our unit being the only paediatric surgical unit on the island.

Congenital anomalies in Bahrain have a different pattern than Europe and North America. We noted three patterns of incidence of congenital anomalies. Some anomalies have the same incidence as in Europe and North America, some anomalies have lesser incidence and some have higher incidence. For example, neonatal intestinal obstruction in Bahrain has almost the same incidence as reported from Liverpool in England. Smithells et al[8] reported the incidence of neonatal intestinal obstruction 15/10,000 live births in Liverpool, and our report shows an incidence of 14/10,000 live births. Cleft lip and palate have an incidence of 15.4/10,000 in Liverpool[8] and 11.5/10,000 in Bahrain. Neural tube defects are far more common in United Kingdom than Bahrain. The incidence of spina bifida cystica and hydrocephalus in United Kingdom varies from 33.6/10,000 in Liverpool to 102/10,000 in Belfast[1], while in Bahrain it is 12.3/10,000. Another example is the incidence of anorectal malformation which is higher in Bahrain than Europe. The incidence in Britain is 2/10,000, in France it is 3/10,000 while in Bahrain it is 4.5/10,000[2]. King[8] reported the possible incidence of posterior urethral valves in Europe and North America to be 2/10,000 males live births which is less than one half the incidence in Bahrain

(2.3/10,000 live births and 4.6/10,000 male live births).

Previous attempt was made to study the incidence of congenital anomalies in Bahrain at neonatal period[9]. Our attempt was concentrated on studying the incidence of the surgical correctable congenital anomalies which reached to our unit at neonatal period or later stage of infancy or childhood. In our study many of the cases which have been missed at neonatal period were diagnosed at later stage. There are many limitations for studying the incidence of congenital anomalies, one of the major limitation is the misdiagnosis of many congenital anomalies which might not be detected at birth or at later stage of childhood. We believe more careful screening for congenital anomalies, national registry for congenital anomalies and combined efforts from the specialities concerned can produce more accurate incidence of various congenital anomalies in Bahrain.

CONCLUSION

There are three patterns for the incidence of correctable congenital anomalies in Bahrain. Most of these anomalies have the same incidence as in Europe and United States, some are more common (anorectal malformations, and posterior urethral valve), and few are less common (myelomeningocele and hydrocephalus).

REFERENCES

1. Rasromani KH, Mohammed A. Pediatric surgical conditions in Bahrian. Presented to the First Congress of Pediatric surgery in the Arabian Peninsula, Riyadh, Saudi Arabia, November 1987.

2. Smithells RW. Incidence and causation of congenital defects. In: Rickham P, Lister J, Irving I. 2nd ed. Neonatal Surgery (Chap 1) London: Butterworth, 1978: 3-9.

3. Stevenson AC, Johnston HA. Congenital Malformations. A Report of a Study of Series of Consecutive Births in 24 Centres. Bull Wld Hlth Org 1966:(suppl 34).

4. Neel JV. A study of major congenital defects in Japanese children. Am J Hum Genet 1958;10:398.

5. Rickam PP, Mawdsley T. The effect of early operation on the survival of spina bifida cystica. Develop Med Child Neurol. 1966;(Suppl 11): 20.

6. Leck I. The geographical distribution of neural tube defect and oral clefts. Br Med Bull 1984;40:390-5.

7. Rasromani KH, Young DG. Current experience with neural tube defects in the west of Scotland. Kinderchirugrie 1979;4:307-14.

8. Smithells RW. Incidence of congenital anomalies in Liverpool. Br J Prev Soc Med 1968;22:36.

9. King LR. Posterior urethral valve. In: Kelalis P, King L, Belman A. 2nd ed. Clinical Pediatric Urology: (Chap 16) Philadelphia: WB Saunders, 1985; 528-54.

10. Al Arrayed S. Congenital anomalies in Bahrain. BMB 1987;9:70-73.