ORIGINAL ARTICLE

The Incidence of Congenital Malformation in the Gadjah Mada University Hospital Yogyakarta During 1974—1979

by

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Abstract

A study of the incidence of congenital malformation among 4625 newborns in the neonatal unit, Gajah Mada University Hospital Yogyakarta, during a period of 5 years (1974 — 1979) is presented. The incidence of 1.64% out of 4625 newborns was found.

The three most frequent malformations were hydrocephalus (21%), cleft lip and cleft palate (9,2%) and Down's Syndrome (9,2%).

A total of 40% were found in multiparous mothers between 20 — 34 years of age, and of 26% in grandemultiparous mothers older than 35 years.

According to Arey's distribution of congenital anomalies, developmental arrest was most prevalent (59,2%).

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Introduction

Data on the incidence of congenital malformations vary greatly. Infants with abnormalities varied from 0,75 to 1,98% according to official records and birth certificates (Mc Keown and Record, 1960). These estimates were rather low compared with data from hospital and clinic birth records in which a variation of 1,43 to 3,3% was noted (Stevenson et al., 1950). Different procedures of investigation for establishing the final diagnosis of congenital defect (Suharyono et al., 1969, Justin Simatupang et al., 1977), and racial varieties may also influence the figures, since frequency and types of malformations vary from race to race (Stevenson et al., 1950).

Despite rapid development in the field of teratology, knowledge of congenital malformations in humans has increased relatively little. At present approximately 10% of all known human malformations are caused by environmental factors and another 10% by genetic and chromosomal factors; the remaining 80% are presumably caused by the intricate interplay of several genetic and environmental factors (Langman, 1975).

The frequency with respect to organ systems is as follows : brain and spinal cord (60%); stomach and intestines (15%); heart and blood vessels (10%); bones, muscles and skin (10%); all other organs (5%) (Arey, 1963).

In Indonesia at this time no data are available on birth defects, neither at national level nor in Yogyakarta.

The aim of this study therefore is to obtain data on newborns with congenital anomalies born in the Gajah Mada University Hospital, Yogyakarta.

Material and methods

The material consists of 4625 newborn infants, in the neonatal unit, Gajah Mada University Hospital, Yogyakarta, during a 5 years period from 1974 - 1979. The distribution of the malformation is determined primarily according to the International Clasification of Diseases (9th revision, 1977). The diagnosis of the malformation is based primarily on physical examination, but if necessary radiological, cardiological, neurological, biological and haematological investigations are carried out. The mothers of these babies were grouped according to parity and age.

TABLE 1 : *Yearly distribution of Congenital Malformation at the Gajah Mada University Hospital (1974 — 1979)*

Year	Number of newborn infants with congenital malformation	Number of newborn infants	Percentage
1974	9	589	1,53%
1975	10	630	1,59%
1976	7	646	1,08%
1977	11	930	1,18%
1978	5	872	0,57%
1979	34	958	3,65%
Total	76	4625	1,64%

TABLE 2 : *Types and number of cases with congenital malformation*

Congenital malformation	Code *)	Number of cases **) in a 5 years period (1974 — 1979)
1. Anencephaly	740.0	3
2. Spina bifida	741	1
3. Encephalocele	742.0	4
4.. Microcephaly	7.42.1	2
5. Hydrocephalus	742.3	16
6. Anophthalmus	743.0	2
7. Microphthalmus	743.1	2
8. Buphthalmus	743.2	2
9. V S D	745.4	4
10. P D A	747.0	2
11. Laryngomalacia	748.3	1
12. Cleft palate	749.0	1
13. Cleft lip	749.1	4
14. Cleft lip and cleft palate	749.2	7
15. Meckel's divertikulum	751.0	3
16. Anal atresia	751.2	5
17. Congenital megacolon	751.3	6
18. Hypospadia	752.5	2
19. Hypoplasia of penis	752.8	2
20. Polydactily	755.0	3
21. Syndactily	755.1	5
22. Phocomelia	755.2	3
23. Down's Syndrome	758.0	7
24. Trisomy 13	758.1	2
25. Conjoined twin	759.4	1

*) In accordance with the numbers in WHO. International Classification of Diseases the 9th revision edition 1977.
**) Some with multiple congenital malformations.

The distribution of congenital malformation of the newborn infant in this study was also determined by Arey's Classification (1963).

Results

Table 1 shows an incidence of 1.64% of congenital malformations in the neonatal unit, Gajah Mada University Hospital, Yogyakarta (76/4625 × 100%) during a 5 years period.

The three most frequent malformations in this study were: Hydrocephalus — 16 cases (21%); Cleft lip and cleft palate — 7 cases (9,2%) and Down's Syndrome — 7 cases (9,2%) (Table 2).

A total of 40.79% of newborns with congenital anomalies were found in multiparous mothers between 20 - 34 years of age, and of 26,36% in grandemultiparous mothers older than 35 years.

Five grandemultiparous mothers with Down's Syndrome baby were older than 35 years, and two multiparous mothers were 20 - 34 years old. (Table 3).

TABLE 3: *Age and parity distribution of the mothers of 76 newborns with congenital malformation during a 5 years period (1974 — 1979)*

Parity \ Age/years	< 19	20 — 34	> 35
Primiparous	3 (3,95%)	13 (17,11%)	0
Multiparous	0	31 (40,79%)	2 (2,63%)
Grandemultiparous	0	7 (9,21%)	20 (26,32%)
Total 76	3	51	22

Discussion

This study shows an incidence of 1,64% of congenital malformations in the neonatal unit, Gajah Mada University Hospital, Yogyakarta during a 5 years period, which is higher than that at Dr. Ciptomangunkusumo General Hospital, Jakarta (0,57%) (Suharjono et al., 1969) and Medan (0,84%) (Justin Simatupang et al., 1977). On the contrary the incidence in our study is approximately similar to that from abroad (1,42 — 3,3%).

It should be noted that there are many apparent discrepancies between the figures reported by different authors, due to the fact that nearly every survey is undertaken for different reasons or with different material or methods (Suharyono et al., 1969; Justin Simatupang et al., 1977).

In our opinion, keeping in mind that in Indonesia the role of the traditional midwife is still significant, i.e. 80 - 90% of births are still attended by traditional midwives (Anna Alisyahbana, 1979), newborns with minor congenital anomalies with no medical or cosmetic consequences (Marden et al., 1964) were never reported to the Health Center or Hospital. On the other hand, newborns with major congenital anomalies which effected seriously both functional and social capacities (Marden et al., 1964) died before being reported to the Health Center or Hospital, making accurate overall estimation of the incidence hardly possible.

The three most frequent malformations in this study were : Hydrocephalus — 16 cases (21%); Cleft lip and cleft palate — 7 cases (9,2%) and Down's Syndrome — 7 cases (9,2%) (Table 2). The incidence of hydrocephalus in our study was lower than at Dr. Ciptomangunkusumo Hospital, Jakarta in 1969 i.e. 34,98% (Suharyono et al., 1969). The figure of hydrocephalus in Medan was 0,8 per thousand in 1977. Arey's figures (1963) were malformations of the brain and spinal cord 60%, and the rest (40%) involving other organs.

The figures of cleft lip and cleft palate in our material are lower than those in Medan (1,7 per thousand of total births). On the other hand figures of Down's Syndrome in our study are higher than those at the Dr. Ciptomangunkusumo Hospital, Jakarta in 1969 (3.5%) and in Medan (0,5 per thousand of total births) in 1977. The findings in this report do not differ very much from figures abroad (1,5 per thousand of births; Bartram 1975).

A total of 40,79% of newborns with congenital anomalies were found in multiparous mothers between 20 - 34 years of age, and of 26,32% in grandemultiparous mothers older than 35 years.

Five grandemultiparous mothers with Down's Syndrome babies were older than 35 years, and two multiparous mothers were 20 - 34 years old (Table 3).

It is well-known that advanced maternal age is mostly one of the reasons for antenatal genetic studies. The chance of a prospective mother, between 40 and 44 years of age, having a child with a chromosomal abnormality is approximately 1 in 40. Available data for women between 35 and 39 years of age reveal a risk of about 2 percent for all chromosomal abnormalities (Milunsky, 1977).

According to Arey (1963) developmental arrest was the most prevalent (59.2%) — Table 4.

Except under controlled experimental conditions, it is difficult or impossible to identify the exact cause that serves a developmental aberration from the normal course.

Any developing organ (or part) passes through an individual critical period (or periods) during which it undergoes accelarated growth and differentiation, and manifests marked susceptibility to injurious influences brought to bear on it.

It is necessary to draw a distinction between genetic constitution (i.e., the genic content of the nuclei) of a given individual and his somatic appearance.

For the genetic constitution, the term "genotype" is applied and for the external appearance the term "phenotype" is used. The phenotype results from the interaction between genes and environment and since the environment can be varied, the phenotype is also potentially capable of variation.

The influence affecting the genotype can be induced by experimental treatments. The potency of radiation in this respect when applied to the mammalian and human embryo at appropriate times is well established. The influences affecting the phenotype are mechanical, radiation, chemical, deficiencies, disease and maternal impressions (Arey, 1963).

TABLE 4 : *Distribution of congenital malformation according to WHO and Arey criteria*

WHO's criteria	Arey's criteria	Number of cases in a 5 years period (1974 — 1979)
1. Anencephaly	Developmental failure	3
2. Anophthalmus	„	2
3. Phocomelia	„	3
4. Spina bifida	Developmental arrest	1
5. Encephalocele	„	4
6. Microcephaly	„	2
7. Hydrocephalus	„	16
8. Microphthalmus	„	2
9. V S D	„	4
10. Cleft palate	„	1
11. Cleft lip	„	4
12. Cleft lip and cleft palate	„	7
13. Hypospadia	„	2
14. Hypoplasia of penis	„	2
15. Buphthalmus	Developmental excess	2
16. Polydactily	„	3
17. Conjoined twin	„	1
18. Syndactily	Failure to subdivide	5
19. P D A	Failure to atrophy	2
20. Meckel's divertikulum	„	3
21. Anal atresia	„	5
22. Laryngomalacia	Atypical differentiation	1
23. Congenital megacolon	„	6
24. Down's Syndrome	„	7
25. Trisomy 13	„	2

REFERENCES

1. ANNA ALISYAHBANA : Masalah perinatologi di masyarakat Indonesia umumnya dan khususnya di daerah penelitian Ujungberung Bandung. Diajukan pada Simposium dan Seminar Perinatologi Nasional II di Semarang, 1979.
2. AREY, L.B. : Textbook and laboratory manual of embryology, 6th ed., (Saunders, Philadelphia, London, 1963).
3. BARTRAM, J.B. : Mental retardation; in Nelson Textbook of Pediatrics, 10th Asian ed., pp. 134 - 136 (Igaku Shoin, Tokyo 1975).
4. JUSTIN SIMATUPANG; NOERSIDA RAID; BISTOK SAING; HELENA SIREGAR : The incidence of congenital malformation in the General Hospital' (RSUPP) Medan, 1970 — 1975. Paediatr. Indones. 17 : 223 (1977).
5. LANGMAN, J. : Congenital Malformation and their causes in Medical Embryology, 3rd Asian ed., p. 109 (Igaku Shoin, Tokyo 1975).
6. MARDEN, P.M.; SMITH, D.W.; Mc DONALD, M.J. : Congenital anomalies in the newborn infant, including minor variations. J. Pediatr. 64 : 357 (1964).
7. Mc KEOWN, T. and RECORD, R.G. : Malformation in population observed for five years after birth; in Wolstenholme and O'Connor Ciba Found Symp. Congenital Malformation, p. 2 (Little Brown, Boston 1960).
8. MILUNSKY, A. : Antenatal diagnosis of genetic disorders in diseases of the newborn, 4th ed., pp. 29 - 32 (Saunders, Philadelphia, London, Toronto 1977).
9. STEVENSON, S.S.; WORCESTER, J.; RICE, R.G. : Six hundred and seventy seven congenitally malformed infants and associated gestational characteristics. I. General considerations. Pediatrics 6 : 37 (1950).
10. SUHARYONO; SUNOTO; SUDIYANTO; M. SUGIONO; SUTEDJO : The Incidence of Congenital Malformation in the Dr. Ciptomangunkusumo General Hospital, Jakarta, 1962 — 1967. Paediatr. Indones. 9 : 9 (1969).
11. W.H.O. : International Classification of Diseases, 9th revision, pp. 417 - 437. (WHO, Geneva 1977).