

# ANENCEPHALY

## (A Study of 118 Cases)

by

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Though there are several reports on anencephaly in western literature, so far no Indian study has been reported to our knowledge. This communication is a report of 118 cases of anencephalus observed at N. W. M. Hospital, Bombay, from January 1958 to December 1965.

### *Incidence*

The incidence of anencephaly as studied by Penrose (1957) was higher in European population than in Asian. In the present series, 118 cases were delivered of anencephalic monsters amongst 84,763 deliveries, giving an incidence of 0.13%. The incidence reported in the literature ranges from 0.01% to 0.67% (Table 1).

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Total percentage of deformed babies during the period of study was 0.64 in 84,763 deliveries. This includes only gross developmental defects apparent on inspection at the time of birth or during the period of stay in the hospital. This is compared with other series in Table II.

Incidence of major congenital malformations at birth is compared with other reports in Table III.

### *The anencephalic monster*

#### *Sex ratio*

Forty-three infants (36.4%) were males, 73 (67.2%) were females and in 3, sex was not recorded, giving male/female ratio as 1:1.6.

#### *Associated malformation*

Six cases had spina bifida while one had meningocele.

#### *Associated complications*

Forty-eight cases had chronic hydramnios and two had acute hydramnios giving incidence of 42.3%. Fifteen cases had evidence of toxæmia. There was no significant increase in previous abortions in women who had anencephalic infants.

TABLE I  
Incidence of anencephaly in various series

Authors	Period	No. of births	Anencephalics	
			No.	%
Hegnauer	1929-41	141,706	117	0.083
Coffey and Jessup	1953-54	12,552	63	0.502
Stevenson	1938-55	30,855	207	0.671
Malpas	1923-32	13,964	44	0.315
McMahon et al	1936-52	168,654	326	0.194
Stevenson et al	1930-41	29,024	67	0.231
Fraser	1950-55	19,839	42	0.211
Searle	1953	8,267	8	0.097
Hsu	1951-53	32,176	18	0.056
Weischen	1955-62	14,834	17	0.115
Present series	1957-65	84,763	118	0.13

TABLE II  
Incidence of deformed infants

Author	Year	Total No. of infants	Deformed
Malpas (England)	1937	13,964	2.11
Neel (Japan)	1948	63,796	3.01
Hegnauer (Germany)	1951	141,706	0.67
Coffey and Jessop (Ireland)	1955	12,552	1.63
Wei and Chen (Taiwan)	1964	14,834	0.87
Present series (Bombay)	1965	84,763	0.64

TABLE III  
Incidence of major congenital malformations, per 1000 births

Type of anomaly	Neel Japan	England McKeawn	Taiwan Wei & Chen	present series
Anencephalus	0.63	1.93	1.15	1.3
Spina bifida and or meningocele	0.26	2.80	0.41	0.87
Hydrocephalus	0.32	1.76	0.48	1.2
Mongolism	0.09	1.11	0.27	0.15
Harelip and or cleft palate	2.78	1.76	1.92	1.2
Club foot	1.10	3.95	0.62	0.69
Miscellaneous	—	—	—	1.59

### Presentation.

Sixty-four per cent of the cases were booked while the rest were emergency admissions.

A woman is more likely to have an anencephalic infant in the first delivery (38.9%) than in any subsequent deliveries, though the highest incidence is in II V parous group (47.5%).

About two-thirds (67.6%) of the patients came with labour pains while the remaining one-third came with variable symptoms like antepartum haemorrhage (9.4%), draining with or without labour pains (6.7%), increasing distention of abdomen (5.08%), persistent breech or transverse lie (4.1%), absent foetal movements (3.4%), bad obstetric history (1.6%) and acute retention of urine (0.8%). Two patients had bad obstetric history, one was a 4th para with 3 still-births while the other patient was 4th para with one still-birth and one craniotomy. Unfortunately she was misdiagnosed and was subjected to elective caesarean section. Out of 50 patients of hydramnios diagnosed clinically, only 6 complained of increasing distention. Only one patient complained of headache and giddiness, out of 15 having hypertension. The antenatal diagnosis was made in a very small number of cases though associated malformations were suspected in cases with hydramnios. About 60% of the cases came at full-term while about 26% came at 8th month or less. Two cases came before sixth month as acute hydramnios. Eighty cases (67.7%) delivered as cephalic presentation. The incidence of deli-

very as face was 5.9%, while breech had a high incidence of 17.7%.

### Outcome of labour

The termination of pregnancy was either by spontaneous delivery or by interference. The mode of delivery was studied in detail (Table IV).

TABLE IV  
Mode of delivery

Spontaneous delivery	83.9%
A.R.M. - S.D.	4.9%
A.R.M.-Pitocin drip - S.D.	4.9%
Hind water rupture - S.D.	3.3%
Abdominal tapping - S.D.	1.2%
L.S.C.S.	0.8%

SD—spontaneous delivery.

About 84% had spontaneous delivery without any interference. But ten patients required manual removal of placenta and four patients had postpartum haemorrhage. All cases who had artificial rupture of either fore or hind waters, or abdominal tapping, were cases of hydramnios. In a few cases the delivery was hastened by pitocin drip. About 14% required draining of excessive liquor to expedite the process of labour. One patient had unfortunately undergone caesarean section (vide supra). Prophylactic use of ergot reduced the incidence of postpartum haemorrhage.

The weight of the new-born varied from less than 1.8 lbs. to 6.2 lbs. (Table V). Maximum number of infants were less than 2.8 lbs. amounting to 65.1%. Only about 11% were more than 4.9 lbs. One hundred and eleven (94.0%) were still-born while 3 lived for 5 to 20 minutes. The maximum survival was of 5 days.

TABLE V  
Distribution of weight

Wt. in lbs.	No. of cases
1.8 and less	19.4%
1.9 - 2.8	45.7%
2.9 - 3.8	70.1%
3.9 - 4.8	8.4%
4.9 - 5.8	8.4%
5.9 and more	3.3%
Unknown	4.2%

### Discussion

The incidence of anencephalus in our series is lower than that reported in Western literature and is comparable with Asian and African series (Searle, 1953, Hsu, 1956, Wei and Chen, 1965). According to Murphy (1963) more than 80% of the deformed babies can be diagnosed on inspection while Potter (1963) stated that one-third of them would be missed unless autopsy was performed. This may account for overall lower incidence of congenital malformations in Indian babies as compared to reports from abroad. The incidence of anencephalus amongst various malformations has been the highest in the present study, which again does not conform with reports from abroad. It remains to be proved whether the differences in the incidence of congenital malformations have any geographical basis since Searle (1953) has noted varied incidence in different communities living in Singapore. This can only be answered when the definitive teratogenic influences operating in the genesis of anencephaly have been elucidated.

In conformity with our findings, it is reported by Evans *et al* (1963) that 85% of all patients were less than third parae. Thirteen out of their 17 cases were primigravidae. However, Penrose (1957) and Wei and Chen (1965) have not observed the same. Though it is believed that babies conceived during summer months have higher incidence of anencephaly, we have not found any seasonal variation which corresponds with the finding of MacMahon *et al* (1954). Cases have been reported of having four successive anencephalic infants but the present study did not reveal any of the women having more than one anencephalous baby.

Sex incidence of anencephalic infants is quoted as 70% for females (Eastman, 1961) which is in conformity with our findings. No female preponderance was found in the series of Wei *et al* (1965). Hellman (1961) by sex determination of abortions came to the conclusion that a large number of male anencephalic infants died in early stages of pregnancy.

Hydramnios, though the occurrence is variable, is frequently associated, as found in our study. The highest incidence so far reported to our knowledge has been by Labrium and Wood (1961) as 72.8% while the lowest (33%) was reported by Wacker (1963).

Since the clinical features which might lead to a definitive diagnosis of the condition occur infrequently, the diagnosis is difficult. However, evidence of hydramnios and abnormal lie should be noted as suspicious. Most of the cases are likely to have

spontaneous delivery. Since induction cannot always be relied upon and in case of failure may make caesarean section imperative, it is not recommended as a routine. Prophylactic use of ergot reduced the incidence of postpartum haemorrhage for which these patients need to be carefully observed.

Live birth of the deformed infant was in conformity with the customary figure of 6 to 8 per cent. In view of the maximum survival reported so far as 2 days (Wei and Chen, 1965) survival of one infant upto five days in the present series is exceptional.

### Summary

One hundred and eighteen cases of anencephaly are studied. The incidence is 0.13% in 84,763 deliveries. Total percentage of deformed babies during the period of study was 0.64%. No seasonal variation was found. Male to female ratio was 1: 1.6. The maximum number of cases was between second to fifth parous groups but primiparae had the highest incidence; 42.3% of cases had associated chronic or acute hydramnios.

The presenting symptom in two-third of the cases was labour pains. The antenatal diagnosis was made in a very small number of cases though 64% of cases were booked. There was high incidence of malpresentation. Most of the cases had spontaneous delivery. The incidence of still-birth was 94%.

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