

## ANENCEPHALUS

### A Study of 40 Cases

by

A. PRABHU,\* M.D., D.G.O.

and

Y. PINTO ROSARIO,\*\* M.D.

An antenatal diagnosis of hydramnios and the presence of a foetal anomaly, especially anencephalus gives a choice of either leaving the patient alone or terminating a useless pregnancy. The behaviour of 40 cases of anencephaly were studied at the Lady Hardinge Medical College, New Delhi, through a period of 3 years from January, 1968 to December, 1970.

The etiology of this condition is not known. According to Sarma (1963) and Corbett (1953) the incidence is not affected by geography, parity and maternal age and no environmental factor can be held responsible for its incidence. There is a strong suggestion that a genetic factor is involved (Eastman 1966). Most probably a hereditary or genetic malformation is transmitted as a recessive gene.

Horne (1958) has reported cases in which 4 successive anencephalic infants were produced by the same parents. Eastman (1966) on the other hand says that the reported geographical differences in the incidences of anencephalus have led to the belief that different environmental conditions in these areas, notably poor living conditions and inadequate diets, predispose to anencephalus. Anderson, Baird and Thomson (1958), in Scot-

land, believe that as social status falls there is a steady increase in the incidence of anencephalus.

Guthlekh (1962) found that anencephalus is 50% more common in the winter months. In this series the majority—55% of patients delivered during the winter months and the next large group 25% delivered in the rainy season between July and September.

The total number of deliveries during this period was 18,348, thereby giving an incidence 0.76% of anencephalus as against the incidence in the following series. Sarma 0.0975%, Vyas 0.13% (1967), Hurwitz (1955) 0.28%, Mahfouz (1949) 0.08%, Book and Rayner (1950) 0.064%. According to Sarma (1963) the total incidence of anencephalus does not appear to be lower in India, while according to Vyas (1967) the incidence is lower in India than in the West. Eastman has noted that 70% of anencephalic monsters are female, while Sarma gives a figure of 75%. In this series, 62.5% were females.

The youngest mother in this series was 19 years and oldest 39 years, the majority being between 20-29 years. In Sarma's series the average age of the mother was 27.6 years. The average parity was 3.4, similar to 3.8 in Sarma's series, while Vyas (1967) found the highest incidence in those who had had 2-5 children.

\*Registrar.

\*\*Professor, Dept. of Gynec. & Obst., Lady Hardinge Hospital, New Delhi.



Other abnormalities are sometimes found in addition to anencephalus, the commonest being spina bifida, talipes equinovarus and umbilical hernia. Malpas (cited by Sarma) found female anencephalics with spina bifida to be twice as common as in male anencephalics with this defect. According to Potter (1952) it is rare to find skeletal defects in association with anencephalus and accessory digits are never present. In this series 4 babies had spina bifida and one had meningocele. Occasionally, twin pregnancies are encountered in which both foetii are anencephalic as was found in one case which was diagnosed radiologically. This patient did not wait for delivery and left hospital against advice. Poly-hydramnios is commonly associated with anencephalus and usually manifests itself after the 28th week of pregnancy.

In a study of 67 anencephalic babies Book and Ryner (1950) recorded a 69% incidence of polyhydramnios. Labrium and Wood (1961) found the incidence to be 72.8%, while in Sarma's series of 128 cases, 36 patients (28%) were associated with a clinically obvious degree of hydramnios, 92 cases being admitted with rupture of membranes and well advanced labour. In this series, 30 i.e. 75% cases had hydramnios and the rest were admitted after rupture of membranes.

Anaemia and pre-eclampsia have been found associated with anencephalus but do not play a significant role. In this series 10% of patients had pre-eclampsia.

There is a definite association of anencephaly with prolonged gestation. In 44 cases in Sarma's series postmaturity was observed in 9 instances. In 2 of these cases, pregnancy went beyond term for 32 and 61 days, respectively. Higgins (cited by Sarma) (1970) recorded a case in which pregnancy went beyond term

for 24 days with foetal movements present. In this series, 5 patients i.e. 12.5% were postmature and had to be induced, but the other patients either came in labour or as soon as they were discovered, they were admitted and induced. Malpas (1951) says that the incidence of postmaturity where the child is anencephalic is high if there is no coincidence of hydramnios. In 1933 Malpas had 44 cases of anencephalus with postmaturity in 9, and in one the pregnancy lasted 374 days.

*Management:* As the malformation is incompatible with life it is better to terminate the unnecessary pregnancy at an early date. As such antenatal diagnosis is important. Antenatal diagnosis was laid down in 1889 by Negri who noted that "by careful palpation the faulty character of the cranium was recognised and there were conclusive movements of the presenting part and limbs".

The diagnosis of anencephalus can be made by finding:

—Permanent exaggerated tension of uterus, with difficulty in finding foetal poles, and in identifying the foetal head. Convulsive movements of foetus and weak foetal heart sounds are additional signs.

The most favourable time for the physical diagnosis of anencephalus is at the beginning of labour, by digital exploration. The form and shape may sometimes permit recognition of an anencephalus when the cephalic extremity presents. The diagnosis becomes easier as labour proceeds. Suggestive signs are small size of the presenting head and absolute absence of the cranial aspect. Hydramnios was present in 75% cases and comparable to Book and Rayner (1950) who gave an incidence of 69%. Difficulty in assessing the presenting part was encountered in 55% cases and diagnosis was



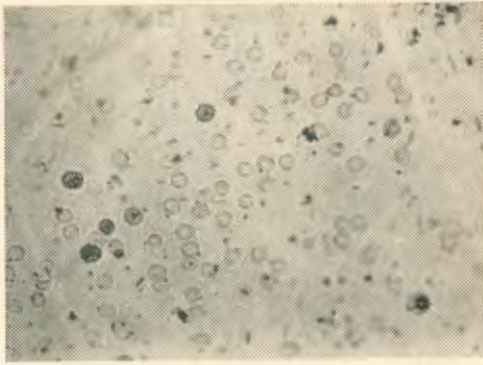


Fig. 1  
Foetal erythrocytes in maternal circulation.

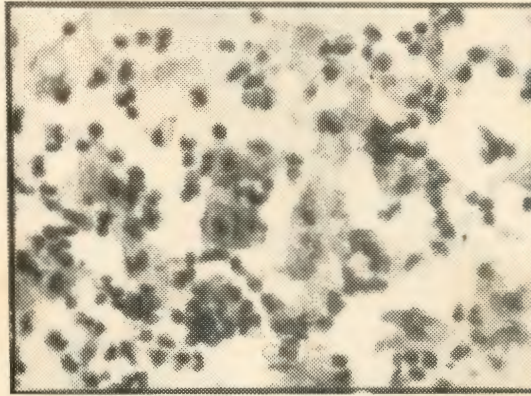


Fig. 1  
Showing perinuclear halo and cannon balls.

Chorioangioma of the Placenta—Banerjee pp. 283-284

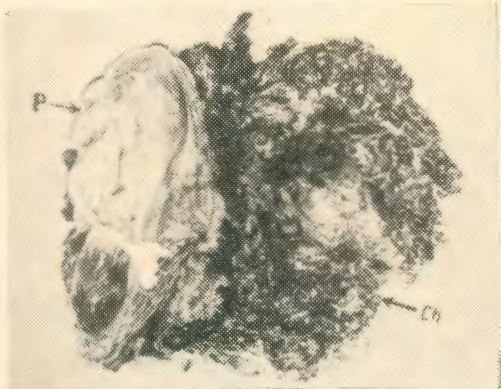


Fig. 1  
Chorio-angioma of the placenta. A dark red mass of enormous size is loosely attached to the maternal surface of the placenta. The mass is divided into lobules of various size and shape.  
(P—Placenta, Ch—Chorioangioma).

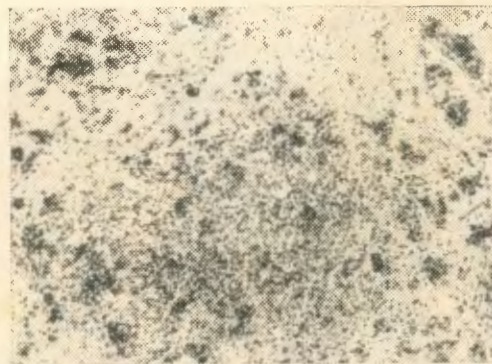


Fig. 2  
Histology shows picture of a tumour closely packed with small blood vessels and capillaries. These are dilated and filled with red blood corpuscles, and are supported by loose net work of connective tissue and stromal cells.





Fig. 1. Case No. 1

5 years after removal of male gonads along with plastic surgery of external genitalia, followed by prolonged treatment with oestrogens.

Fig. 2. Case No. 2 & 3.

Male body contour, male distribution of hair and slight enlargement of the breasts. Brought up as a girl, as shown by bangles around the wrists.

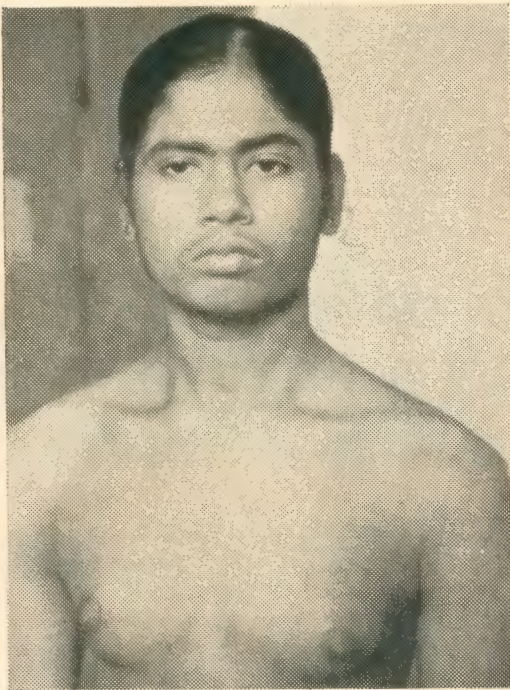


Fig. 3.

Male body contour, male distribution of hair and slight enlargement of the breasts. Brought up as a girl.



Fig. 4. Case No. 2

Appearance of the external genitalia with an enlarged phallus.





Fig. 5. Case No. 2  
External genitalia look more like a bifid scrotum on the right side.

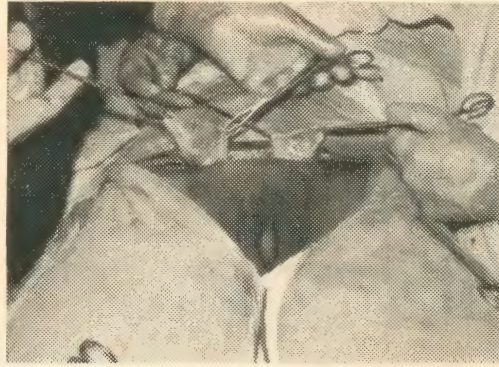


Fig. 6. Case No. 2  
Both inguinal testes exposed during operation.

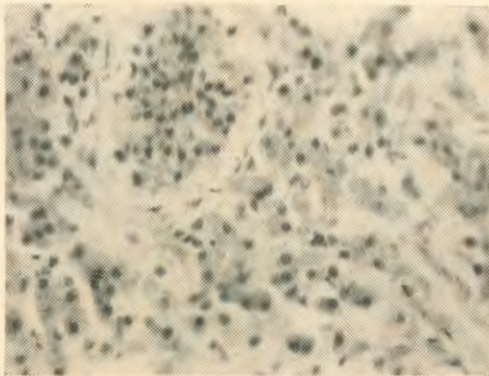


Fig. 7. Case No. 2  
Histological appearance of a gonad; testicular tubules and interstitial cells clearly seen.



Fig. 8.  
External appearance of the youngest normal sister of Case No. 1 and 2.

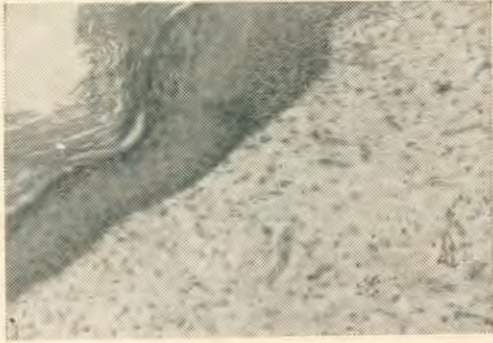


Fig. 1

Microsection of the cyst wall covered by stratified squamous epithelium which shows acanthosis, hyperkeratosis with flattening of rete pegs.

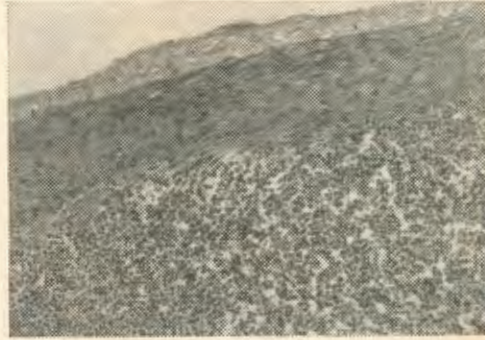


Fig. 2

Section showing subepidermal tissue with fibrocollagenous tissue infiltrated diffusely by chronic inflammatory cells, mostly lymphocytes, round cells and plasma cells.

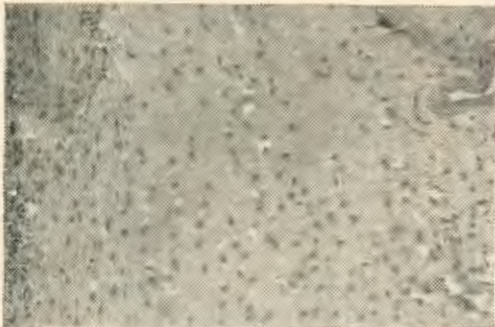


Fig. 3

Cyst wall showing cholesterol clefts, a few pseudoxanthoma cells and fat cells.

Endometrial Sarcoma—Reddy et al pp. 290-293



Fig. 1

Uterus of Case 1, showing a polypoid tumour in the endometrial cavity, occupying 3/4th of the uterine cavity. The myometrium is thinned out.



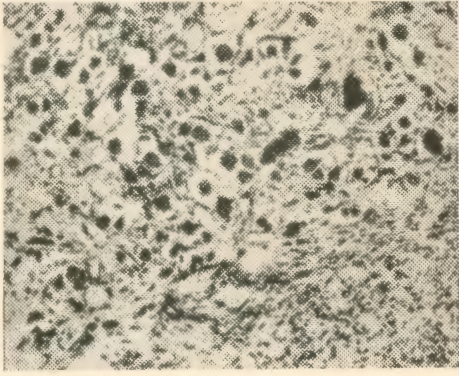


Fig. 2

Photomicrograph of the tumour showing sheets of large polygonal cells with centrally placed nucleus. H. & E. x 400.

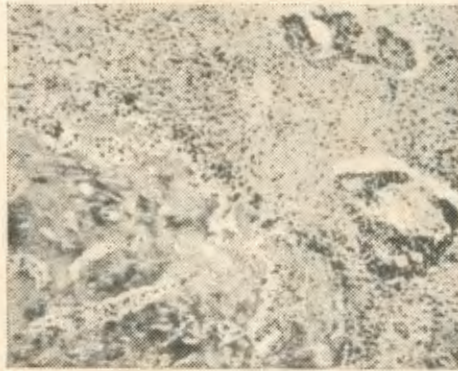


Fig. 3

Photomicrograph of tumour showing areas of immature cartilage.



Fig. 4

Uterus of case 2 showing similar features, as that of case 1.

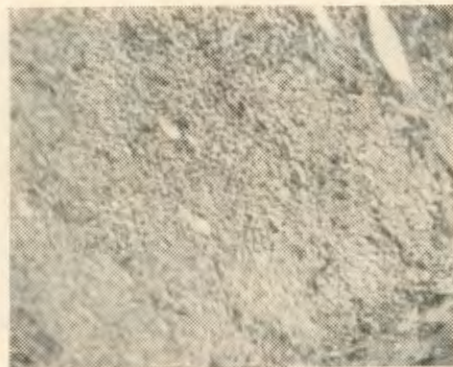


Fig. 5

Photomicrograph of tumours of case 2, showing diffuse sheets of spindle shaped, or fusiform cells with large plump nuclei.



Fig. 6

Photomicrograph showing the neoplastic cells arranged in irregular gland like structures infiltrating the myometrium.



Fig. 7

Showing both carcinomatous and sarcomatous pattern in the same area.

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*Lipoma of Corpus and Cervix Uteri—Hinge et al pp. 294-296*

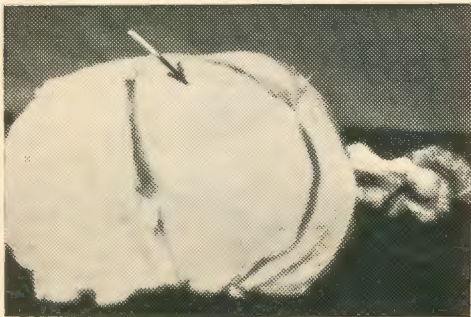


Fig. 1

Cut surface of uterus showing a well circumscribed tumour mass 5 x 4 cms., yellowish in colour. By the side of tumour a small fibroid 3 x 2 cms. cut section whorled, whitish in colour.

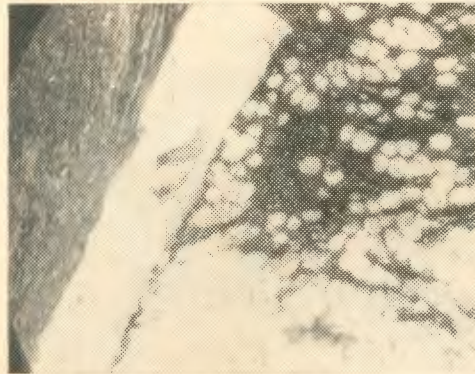


Fig. 2

Photomicrograph showing areas of mature adipose and fibrous septa. Lower down uterine tissue i.e. myometrial and endometrial glands seen (H & E) x 100.



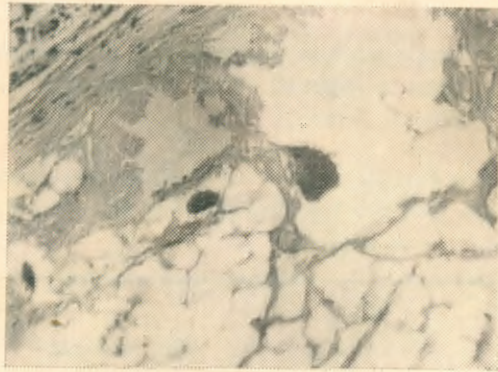


Fig. 3  
Photomicrograph showing areas of mature adipose and cervical tissue with necrosis and calcification (H & E) x 100.



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made by X-ray examination or after rupture of membranes and vaginal examination. Excessive foetal movements was noticed in 12.5% cases. In 65% the presentation was not made out due to the tenseness of the abdomen, while 6 patients (15%) presented as vertex and another 15% as breech and 5% as transverse. There was one case of twin pregnancy which was only diagnosed by X-ray.

After the diagnosis was established antenatally, 20% of patients were induced by an artificial rupture of forewaters and a drip with 2.5 units of syntocinon. Two cases had a syntocinon drip prior to surgical induction. In 10% of cases the membranes had ruptured outside and were delivered by the help of a syntocinon drip. Another 10% had an abdominal amniocentesis and 2.5% cases had a high rupture of membranes and in both instances syntocinon drip was given. Spontaneous delivery occurred in 50% of cases and 2.5% left hospital undelivered.

The duration of labour as compared with the duration of gestation did not show any marked increase or decrease. Most deliveries were completed within 8 hours, a finding comparable to Sarma's series. Three deliveries lasted longer than 24 hours but were complete within 72 hours. The induction-delivery interval studied by Russel and Abbas (1954) showed that delivery was complete in 72 hours in all cases except 6 of 93 cases induced surgically and no harm resulted from delay because all were delivered within a further 18 hours. In relation to period of amenorrhoea at the onset of labour they proved that there was a higher incidence of delay in the induction-delivery interval when the amenorrhoea was under 32 weeks.

In a large number of cases the foetus may be born spontaneously, but in some

instances the baby reaches an appreciable size and the shoulders tend to be excessively large and well formed, and this may produce such a measure of disproportion which in turn can give rise to labour dystocia. Impaction of the shoulders when the anencephalic monster is of appreciable size is a complication which sometimes requires complex and difficult obstetric manoeuvres to effect delivery. The broad shoulders often become arrested at the brim or in the midcavity. In Sarma's series cleidotomy was done in 3 cases, while two cases had constriction ring dystocia and rupture of uterus in labour. There were no such cases in this series.

A complication which should be guarded against is the remote possibility of precipitating placental detachment in a polyhydramnios patient by rupturing the membranes and releasing the fluid suddenly. Gough (1959) suggested the sudden decrease in the internal surface area of the uterus following rupture of membranes in hydramnios is the cause of placental separation as occurred in 8 of his cases. He advocated trans-abdominal amniotomy with slow withdrawal of liquor as a means of reducing this danger. In this series 4 cases of abruptio placentae occurred and the amount of retroplacental clots formed measured from 4 to 14 ounces after rupture of membranes.

Russell and Abbas (1954) studied a selected series of 153 cases of anencephalus with hydramnios and found no difference in antepartum haemorrhage rate between a group of 93 in whom labour was induced. However, they give an incidence of 10.4 per cent of cases in whom accidental haemorrhage was associated with anencephaly. Jones (1967) gives an incidence of 1.4 per cent.



In this series only 3 babies were born alive, 2 died immediately after birth and one baby lived for 5 hours. Live births found by Vyas (1967) were 6.8% and one infant survived upto 5 days. The maximum reported survival otherwise is 2 days (Wei and Chen, 1965).

#### Conclusion

Forty cases of anencephaly were studied, 55% of them delivered in the winter months, females predominated (62.5%). 75% of cases were with associated hydramnios. Only 12.5% were post-mature. Labour was induced in 20% of cases with a syntocinon drip following artificial rupture of forewaters. Spontaneous delivery occurred in 50% of cases. There was no incidence of prolonged or obstructed labour.

#### Acknowledgement

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