ROLE OF CONGENITAL ANOMALIES IN PERINATAL MORTALITY†

by

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Introduction

The perinatal mortality serves as a sensitive index of maternal and neonatal care for an area and it also reflects the general health and other important socio-biological features of mother and infant in that area. Survey of medical literature reveals the stupendous waste of life in the perinatal period, particularly in the economically backward areas of the world. Perinatal loss, a problem of multiple causes can be assessed and solved to a great extent by a coordinated and intensive effort by obstetrician, paediatrician, pathologist and radiologist. Perinatal deaths need detailed systemic and autopsy studies to assess the relative importance of various causative factors. The aim of the present study is to analyse and assess the contribution of congenital malformations to perinatal mortality.

Material and Methods

The present study has been taken up at the upgraded institute of obstetrics and gynaecology, Government Maternity Hospital, Hyderabad on 6375 consective births over a period of 8 months from January, 1978 to August, 1978. Out of 457 perinatal deaths during the period of study, detailed autopsy study was done on 220 cases with an autopsy rate of 46%, the higest reported so far in such short span of study.

TABLE I
Incidence of Parenteral Mortality

Total deliveries conducted during study period	6,375
Total Perinatal deaths	457
Perinatal mortality rate	71.68/ thousand births
No. of necrosis done	220
Autopsy rate	46%

Observations

There were 24 cases of congenital malformations detected on external clinical examination alone. During detailed

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necropsy, there were 26 more cases of congenital anomalies detected, clearly demonstrationg the value of autopsy study in detecting exact cause of death in clinically undiagnosed cases. Thirty-nine babies had major type of malformation. Among the major type of abnormalities neural tube defects topped the list responsible for 53.5% followed by gastro-intestinal system 15%, respiratory system, 7.5% and genitourinary system 7.5%. As shown in Table II relationship of possible etiological factors could be made in 12 (24%) of cases.

TABLE II

Relationship of Possible Etiological Factors

Condition	No. of cases	Consanguinity	Fever in first trimester	Irradiation	Others
Anencephaly	14	2	2	—	—
Hydrocephalous	5	4	—	—	—
Thanatopheric Dwarfism	1	—	—	6 months exposure to irradiation at Nuclear Fuel Complex	—
Meconium Peritonitis	1	—	—	—	History of Infective Hepatitis in first Trimester
Exemphalous	2	1	1	—	—

Details of some interesting congenital anomalies seen during period of study.

Anencephaly

There were 14 cases of anencephaly with well marked female predominance in ratio of 11:3. Hydramnios was associated with 65% of cases. All cases showed enlarged thymus. Adrenal hypoplasia was a universal phenomenon with leafy thin glands with combined weight less than 1 gram. (Photo I with legend).

Tracheo-oesophageal Fistula

There were 2 cases of tracheo-esophageal fistula both had hydroamnios delivered following premature labour. Both were vigorous at birth (as against R.D.S. with which this condition is often confused, where Apgar scoring was about 5-6). After being vigorous at birth, an hour or two later they deteriorated by increase in respiratory rate and peripheral cyanosis like hyaline membrane disease. They showed little response to routine revival attempts by O_2 inhalation, airway, suction and I.V. $NaHCO_3$ and glucose. The diagnosis is pointed by failure to pass Naso-gastic tube, and is confirmed by X-ray. On autopsy, one of our cases showed pulmonary oedema and the other revealed severe bilateral bronchopneumonea to which such babies are predisposed (Photo 2 with legend).

Diaphragmatic Hernia

In course of study, 4 cases of diaphragmatic hernia were seen, 3 of them showing defect in left dome and in 1 almost complete absence of right dome. The babies were vigorous at birth but started deteriorating within 5-10 mts. and showed little improvement to resuscitatory efforts with O_2, airway suction and I.V. $NaHCO_3$ and glucose administration. Diagnosis is not difficult provided the possibility is

kept in mind by auscultatory finding of dextrocardia, intestinal sounds in thoracic cage and followed by radiological confirmation, central cyanosis and deterioration of neonate is due to mechanical embarassment of abdominal contents on heart and lungs. (Photo 3 with legend).

Cloacal Anomaly

This premature baby had intrapartum death following soft tissue dystocia showing irregularly distended abdomen. Autopsy revealed descending colon continued as a large cystic swelling, thin walled ending blindly at the level of left iliac fossa without any evidence of rectosigmoid or prostate. Apparently normal looking penis was without urethera in penile shaft. Thus there was non-differentiation of rectum, anal canal, urinary bladder and urethra—consistent with absence of cloacal differentiation (Photo 4 with legend).

Urachal Cyst

A multipara in premature labour had intrapartum death from soft tissue dystocia. Abdomen was grossly distended and soft in consistency. A multiloculated cyst with engorged veins was anteriorly seen with undifferenciated external genitalia extending between bladder and umbilicus. (Photo 5 with legend).

Posterior Urethral Valve

A primipara, 30-32 weeks pregnant had premature labour with oblique lie. X-Ray revealed soft tissue abdominal opacity with pressure effects noted on the vertabral column and ribs (crowding of upper ribs and flaring of lower ribs). About 1500 ml. of fluid was tapped from distended abdomen and doubled up still born foetus was delivered. There were multiple haemorrages in anterior abdominal wall. On autopsy, what was apparently looking like fetal ascites was an enormously distended bladder with bilateral hydroureter from valvular obstruction in posterior urethra. (Photo 6 with legend).

Osteogenesis Imperfecta

Antenatal diagnosis is difficult because of poor visualisation from associated hydramnios. The baby is short stumpy with limb deformities. In our case it was thick bone type which can be diagnosed prenatally and is invariably fatal. The others are slender and cystic varieties, limb bones are short stumpy, broadened and deformed at the ends due to multiple fractures with excess callous formation. Ribs are similarly affected showing beaded appearance. Vertebral column is normal in our case. Cranial vault is not visualised in some areas. There are non-calcified areas sorrounding ossified areas giving the skull a mosaic appearance. (Photo 7 with legend).

Conclusions

1. Congenital anomalies was No. 2 cause of parinatal mortality (17.8% of P.N.M.).

2. Neural tube defects topped the list, responsible for 53.5% of congenital anomalies.

3. Possible factors predisposing to malaformations have been highlighted.

4. The value of autopsy in pointing out exact congenital malformation and in

detecting exact cause of death in clinically undiagnosed or suspected cases has been clearly demonstrated.

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See Figs. on Art Paper I-II