pathology in our patient was characteristic of the classical variety.

The ideal mode of management is still uncertain. The first line of management in the cases reported, is surgery-either total or partial excision of the tumor mass. Twenty eight of the 35 cases in Rubinstein's series underwent surgery of whom 22 patients survived post operation. Nine of these patients had recurrences in a period ranging from 6 months to 7 years.

All 11 cases in Berger's series were operated and were also subjected to post-operative radiotherapy to prevent recurrences. Four patients showed a recurrence. Recurrences are reported to be common in those patients with solid tumors and those undergoing subtotal resection. Recurrences were managed by surgery or a combination of multiple chemotherapy with radiotherapy.

In summary, primary cerebral neuroblastoma is a rare type of neuroectodcural tumor. Probably this is the first case reported, that has presented at birth.

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Post Mortem Radiography of Perinatal Deaths: An Aid to Genetic Counselling

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Post mortem radiography of perinatal deaths is a simple and informative investigation which, in selected cases, can help in reaching a correct diagnosis and accurate genetic counselling thereafter(l).

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In our country perinatal autopsy is not a routine because of constraints in the form of expertise, finances, time, space and social issues. As a result diagnosis remains doubtful; hence a correct recurrence risk and possibility of prenatal diagnosis cannot be communicated to the parents. Post mortem radiography of selected cases can be a positive step in this direction.

We present our experience of this technique and outline some gross signs which can help in selection of perinatal deaths for radiography. .

Material and Methods

Over the past one and a half years we have performed 50 fetal autopsies. The cases were primarily referred by the obstetricians because of congenital malformations. Each fetus was dealt with according to established protocol in the order mentioned, *i.e.,* : *(i)* Photography; *(ii)* Gross examination; *(iii)* Anthropometry; *(iv)* Radiography consisting of an AP and lateral view of the whole fetus; *(v)* Autopsy; and *(vi)* Histology of cord, placenta, bone and internal organs. From this group we have selected 6 fetuses whose correct diagnosis could be made on gross examination and radiography. The autopsy did not yield further significant information. These 6 fetuses constitute the subjects of the present report.

Results

Table I shows the gestational age, gross, radiological findings together with the autopsy and histology. We have also mentioned the diagnosis, mode of inheritance and risk of recurrence.

Discussion

The clinical, radiological, autopsy and histological findings of six fetuses are presented. The results have shown that a conclusive diagnosis of all these cases could be made on the basis of gross examination and radiology alone. Although autopsy/ histology (done in 5 cases) did yield further findings it did not add to or change the diagnosis. In Cases I and II there was a diagnostic dilemma since the fetuses had features common to both hydrolethalus and Majewski syndrome. This could not be resolved even by autopsy /histology(2).

It is not, however, implied that fetal autopsy is non essential. On the contrary autopsy of all still borns and neonatal deaths should be a routine(3-4). However, as mentioned earlier there are constraints and adequate facilities are available only at tertiary centres. Fetal radiography on the other hand can be carried out wherever there is an X-ray machine, even at a primary health care centre.

As the data has shown, perinatal radiography is diagnostic of skeletal dysplasias which have an incidence of *9/1000* perinatal deaths(5). The presence of 6 such cases in our series of 50 can be accounted for by a biased referral of fetuses with obvious congenital malformations. A routine perinatal radiography, although desirable, may not be cost effective. Hence, on the basis of this study, some gross clinical signs have been identified which should help in selection of cases for radiography. These features are: hydrops, short limbs, polydactyly, arid small or bell shaped thorax.

Selective radiography of perinatal deaths having one or more of the above mentioned features is likely to result in a higher positive yield and improved genetic counse11ing thereby.

TABLE I–*Radiology and Autopsy Findings Together with Recurrence Risk*

S.No.	Gestation	Gross	Radiology	Diagnosis	Inheritance	Recurrence risk	Autopsy	Histology/ (Bone)
1.	20 weeks	Short limbs large head, cleft lip, polydactyly all 4 limbs micrognathia, small thorax, CTEV	Short ribs, shortened long bones, absent tibiae, normal spine and pelvis	Majewski Syndrome or Hydrolethalus syndrome	Autosomal recessive	25%	Absent corpus callosum, hydrocephalus, absent lobation of lungs.	Growth plate showed irregular chondro-osseous transformation zone. Hypertrophic chondral zones were narrow and consisted of irregular columns and abnormal clusters of cells
2.	21 weeks	All above findings and hydrops	Short ribs, shortened long bones, hypoplastic tibiae. Normal spine and pelvis	Same	Same	25%	Same as above together with ventricular septal defect in heart	Same
3.	36 weeks	Hydrops, polydactyly, short limbs, narrow thorax	Short ribs, flat vertebral bodies, shortened long bones, with irregular margins dysplastic pelvis (*Fig. 1*).	Saldino Noonan syndrome	Autosomal recessive	25%	Autopsy consent witheld	
4.	31 weeks	Short limbs, depressed nasal bridge, narrow thorax, hydrops	Short ribs, flat 'H' shaped vertebral bodies, short bowed long bones with broad and flared metaphyses 'telephone receiver' like femur (*Fig. 2*)	Thanato-phoric Dwarfism	Autosomal dominant	0%	No additional finding	Growth plate showed marked reduction and disorganization. The columns of hyperplastic cartilage are greatly reduced. The resting cartilage appears normal

5.	32 weeks	Short limbs, hydrops, cataract, fragile tissue, narrow thorax.	Defective ossification cation overall, skull not ossified; short and crumpled long bones, beaded ribs, flat vertebrae	Osteogenesis imperfecta Type II	Autosomal dominant or Autosomal recessive	25% consanguinous couples. 4% in non-consanginous	No additional finding	The growth plate, metaphysis and diaphysis cartilage are greatly reduced. The resting cartilage appears normal.
6.	22 weeks	Short 'flipper like' limbs, hydrops, Narrow thorax	Short ribs with splayed ends, vertebrae poorly ossified, long bones are short broad and undermodelled, with cupped metaphysis short irregular ilae	Achondrogenesis. Type-II	Autosomal recessive	25%	No additional finding	Disorganized growth plate with scanty column formation. Resting cartilage was hypercellular with reduced matrix and excess of fibro vascular cones. Occasional swollen chondrocytes with inclusion bodies seen

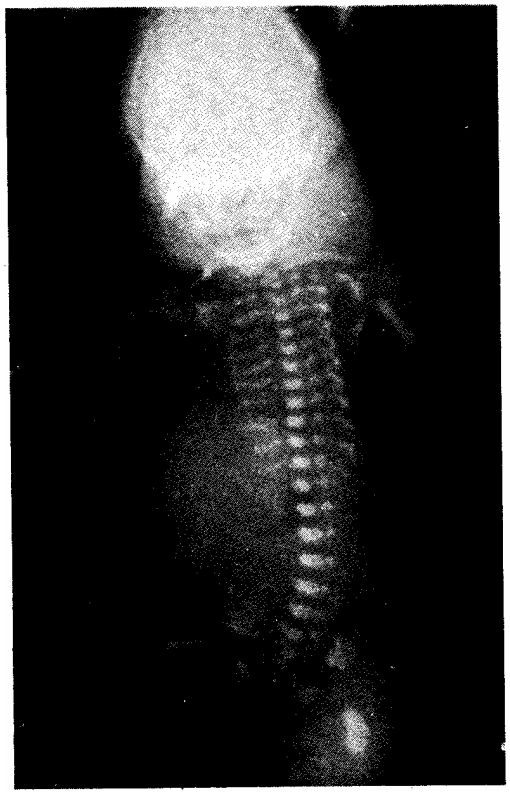

Fig. 1. *Infantogram showing shOI1 ribs, flat vertebrae, shol1ened long bones with ilregular margins and dysplastic pelvis.*

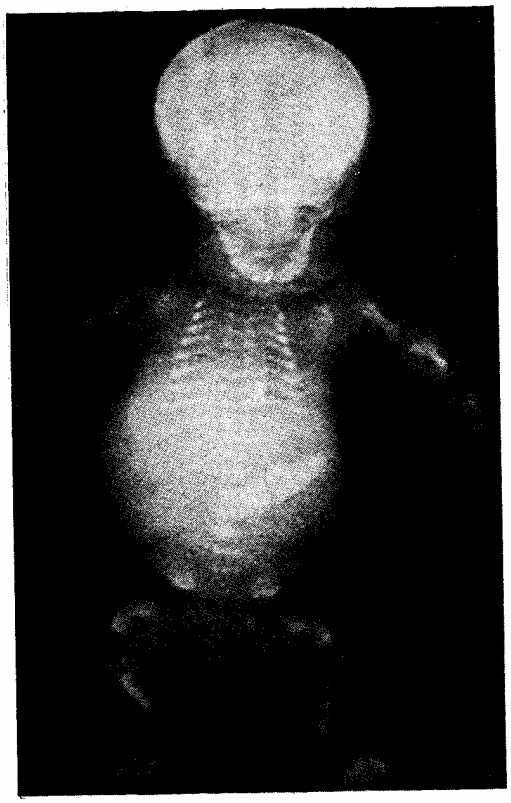

Fig. 2. *Infantogram showing shol1 ribs, flat vertebrae, shol1ened long bones and 'telephone receiver' like femur.*

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