"A CASE OF IDENTICAL HYDROCEPHALIC ACHONDROPLASIC TWINS"

by

S. K. Desai*, M.B.B.S., D.G.O., S. V. Chitnis**, M.B.B.S., D.G.O.
K. M. Masani***, M.D. (Lond.) F.R.C.S., F.I.C.S.

The association of hydrocephalus and achondroplasia has been recognised in the past, but the occurrence of identical hydrocephalic achondroplastic twins is indeed very rare. The record of the past 30 years of this hospital reveals this to be the first such case at the Nowrosjee Wadia Maternity Hospital, Bombay. Scanning through literature the paucity of references to this condition is only too obvious. The first case of identical achondroplasic twins was referred by Goddam in 1945, but it was Sydney Cohn and Weinberg in 1956 who reported a case of identical achondroplasic twins with hydrocephalus.

Case Report

A Hindu patient aged 30 years, (Conf. No. 4163) para V gravida V, registered for confinement at N.W.M. Hospital in the 26th week of her pregnancy. Clinical examination on first antenatal visit revealed a uterus appearing to be of 28th weeks in size. Foetal parts were well felt and foetal heart sounds were regular. There was no evidence of toxaemia or anaemia. Cardiovascular system revealed no abnormality. On examination of the respiratory system crepitations were heard at the right apex. Fluroscopic examination revealed a tuberculous infiltration in right midzone. Hence the patient was treated with antitubercular line of treatment (streptomycin and isonex).

The patient had previous 4 full-term normal deliveries. All the babies were alive and well. Her last confinement was in the year 1961. There was no history of previous exposure to irradiation or teratogenic agents. There was no family history of diabetes or malformation.

On follow up examination in the antenatal clinic patient developed hydramnios at 32 weeks. An x-ray of abdomen was taken, which revealed presence of twins, both presenting as breech; the skull outlines were not well visualised.

At 36 weeks the patient was hospitalised for breathlessness. Abdominal paracentesis was performed and 26 ounces of amniotic fluid was removed to relieve her cardio-respiratory embarassment. Six hours later patient went into spontaneous labour and delivered both the babies as complete breech on 14-6-65 at 5.30 P.M. There was no difficulty in delivery of after-coming heads in either of the babies. The hydrocephalic heads squeezed out slowly as there was hardly any bony development in the skull.

First baby was female weighing 2 kgs. and a fresh still-birth.

The second baby was also a female weighing 2 kgs; It just gasped after delivery and expired in 5 minutes. Placenta was expelled in 10 minutes, there was no post-partum haemorrhage. Examination of placenta showed it to be of uniovular

*Resident Medical Officer.
**Resident Accoucheur.
***Hon. Principal Medical Officer, and Hon. Visiting Obstetrician, Nowrosjee Wadia Maternity Hospital, Bombay 12.
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Fig. 1
Showing identical twins.

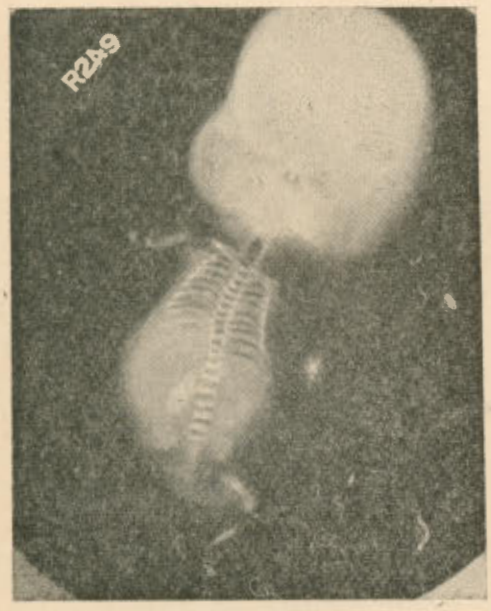

Fig. 2
X-ray of one of the twins. X-ray showing hydrocephalic skull.

variety. Histo-pathological report showed no tubercular infiltration.

Post-mortem examination of babies:

Measurements: length 39 cm., upper extremities 10 cm. trunk 17 cm. lower extremities 10 cm. Chest circumference 29 cm. head circumference 37 cm. Both the babies were hydrocephalic and achondroplasic. Skull bones were not well developed. Other organs were normal. No other malformation was detected.

Comments

Although hydrocephalus in association with achnodroplasia has been described as common by MacCallum and Koller, more recent work has indicated that enlargement of head is due to hypertrophy of bones and often mistaken for hydrocephalus. True hydrocephalus is rare as quoted by Potter. MacCallum ascribed the hydrocephalus to premature synostiosis of portion of sphenoid to one another and to the base of the occiput. Draps stated that the association is exremely rare and the apparent hydrocephalus is due to exaggeration of development of periosteal bones.

In the present case the skull bones were not well developed. They were small and widely separated. The whole of cranial cavity was occupied by cerebro-spinal fluid. When hydrocephalus does occur with chondrodystrophy it has been ascribed to be due to interference with proper circulation of cerebro-spinal fluid. It has been stated that osseous hypertrophy at the foramen magnum may cause sufficient compression to account for neonatal death.

The condition of achondroplasia appears to be inherited as a Mendalian dominant, but the process is complicated by frequent mutation and skipping phenomena. In the above mentioned case there was no family

history of either achondroplasia or hydrocephalus for three generations.

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