Medical Memoranda

Congenital Posterior Pharyngeal Pouch Simulating Oesophageal Atresia

The clinical and radiological signs of oesophageal atresia are well known and the diagnosis is not usually in doubt at the time of operation. In the case described the clinical picture of atresia of the oesophagus was simulated by a pouch of the posterior pharynx.

CASE REPORT

A female child, birth weight 3 lb. (1,360 g.), was admitted at the age of 16 hours to the Nightingale Premature Baby Unit, Derby, on February 13, 1957. Delivery at home had been normal from a pre-eclamptic but previously healthy primipara of 23. The infant had been fed twice before admission, but no regurgitation of feed was reported. When admitted the temperature was 96° F. (35.6° C.), pulse 136, and respirations 48. The colour was good despite a copious flow of mucus from the mouth and pharynx. Numerous rhonchi were heard over both lungs. Mucus was aspirated and no further feeds were given until February 14 (40 hours after birth), when two feeds of glucose-water were given by bottle and retained. Expressed breast milk was then offered, but as it was regurgitated the succeeding three feeds were given by tube; during the third of these difficulty was experienced in passing the tube. At radiological examination a radio-opaque catheter was held up at the level of the second dorsal vertebra. Lipiodol was passed down the catheter and demonstrated a blind upper segment similar to that seen in atresia of the oesophagus.

At operation the oesophagus was normal, and a catheter was passed throughout its entire length. There was no fistulous connexion with the trachea or bronchi. It was then suspected that a pharyngeal pouch was present, and this was confirmed by inserting a smaller "polythene" catheter, which passed directly into the pouch and could be seen protruding down through the thoracic inlet posterior to the oesophagus. As the baby's condition was poor, only gastrostomy could be performed, but the infant died nine hours after operation.

At post-mortem examination a large pouch extended posteriorly behind the oesophagus, the neck of the pouch arising in the posterior wall of the pharynx, opposite the epiglottis. Microscopical examination of the pouch showed the wall to be degenerated and oedematous, with round-cell infiltration. It had a nondescript lining which could have been necrotizing epithelium; such morphological evidence as did occur suggested squamous rather than ciliated epithelium.

COMMENT

Posterior hypopharyngeal diverticula in the newborn are rare, but not unimportant in that they closely simulate atresia of the oesophagus, especially if there is a tracheo-oesophageal fistula. Brintnall and Kridelbaugh (1950) described two such cases that presented with symptoms similar to those of oesophageal atresia. In these cases pharyngoscopy was carried out at operation after a normal oesophagus was found. In the first case the result was inconclusive, as the oesophagoscope passed into the diverticulum and the slit-like oesophageal aperture was thought to be cleft in the oesophageal membrane. In the second case the apertures of the diverticulum and oesophagus were clearly identified. In the case described, presumably the tube passed down the oesophagus normally during some feeds, while at others it was held up in the pouch.

The actual nature of the condition may be reduplication of the foregut or a true diverticulum. True diverticula usually have the same type of mucosal lining as the part of the alimentary tract from which they arise. Gross (1953) states that the epithelial lining of a reduplication resembles some part of the alimentary tract, although this does not

necessarily correspond to the mucosa of the alimentary tract at the level of the reduplication.

It seems probable that these patients will be admitted to hospital as cases of oesophageal atresia, and the diagnosis of pouch of the posterior pharynx made either at pre-operative pharyngoscopy or when a normal oesophagus is found at thoracotomy. The operation of choice in these patients is initially gastrostomy and, if the condition of the child permits, an incision in the right side of the neck to marsupialize the pouch. The excision of the pouch should be carried out later, and it may be advisable to delay this until the child is between 3 and 6 months old.

As an identical clinical picture may be presented by atresia of the oesophagus and posterior pharyngeal pouch, it might be advisable to carry out pre-operative pharyngoscopy in the hope that the opening of a pharyngeal pouch would be seen; and if such an opening were seen, then an attempt should be made to guide a catheter alongside it into the oesophagus.

I am grateful to Dr. B. M. Laurance for permission to publish this case report and for advice in its preparation. I thank Miss B. V. Slessor, who performed the operation, for her advice; Dr. G. Q. Chance for the radiological report; and Dr. D. Barron Cruickshank, Cambridge, for the report on the sections.

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Iris Colour in Mongolism

Deficiencies in pigment production, as manifested by hair colour, have been demonstrated in phenylketonuria (Cowie and Penrose, 1951) and in the infantile Fanconi syndrome (Cowie, 1956). In both these conditions, anomalies in amino-acid metabolism are known to be present. In mongolism, too, anomalies in amino-acid metabolism have been reported, the significance of which remains unexplained (Wright and Fink, 1957). It was therefore thought that it might be useful to investigate pigmentation in mongolism. As the colour of the iris is said to depend partly on the pigment in the stroma cells and partly on that in the cells of the retinal layers (Hine, 1949), it seems reasonable to think that iris colour may reflect disturbance of pigment metabolism.

Further, immature development, producing what Shuttleworth (1909) calls an "unfinished child," is a well-known feature of mongolism. In view, therefore, of Tanner's (1953) statement that, "like the hair, the iris gradually becomes darker as the individual ages except in those with very light eyes to begin with," it was thought that the tendency to immaturity in mongolism might be reflected by a lag of this darkening process in the iris.

Observations on iris colour in mongolism have been recorded (Brushfield, 1924; Engler, 1949). No attempt, however, appears to have been made to compare objectively the iris colour in mongolism with that in a control group.

PRESENT INVESTIGATION

The iris colours of 94 patients with mongolism at the Fountain Hospital, of whom 57 were males and 37 females, ranging in age from 3 to 23 years, were compared with those of an unselected group of mental defectives of the same age and sex from the same hospital.

This was done by means of the "Martin Augen-Farben Tafel." This consists of a metal box in which are embedded a series of 16 glass eyes of varying colours, ranging from dark brown to light blue, and labelled 1 to 16 respectively. On this scale, the lighter the eye the larger the number—thus the darkest eye is labelled 1 and the lightest 16.

The technique used was to record the number of the eye on the scale corresponding closest in colour to the eyes of the subject being examined. The same observer made the

observations throughout and only in natural daylight, keeping the scale in a similar position to that of the eyes of the subject so that the light was impinging directly on them. In this way it is considered that variations due to inconstant light effects were practically eliminated.

While it is recognized that, in the words of Tanner (1953), "the colour of the iris itself is seldom uniform, the general colour being interfered with by superimposed detail colour, usually in the form of spots or small patches," and while it may not always be possible to match a subject's iris colour exactly with one on the scale, any approximation seemed to be close enough to make no significant difference to the overall results. The results are shown in Table I.

TABLE I

Martin Colour Scale :	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16
No. of mongols ..	0	1	3	13	3	5	5	4	3	10	1	9	10	25	1	1
No. of controls ..	0	1	3	16	9	3	6	4	3	3	3	12	3	19	3	6

TABLE II

Iris Shade :	Brown	Blue	Total
Martin Colour Scale :	1 to 8	9 to 16	
Mongols ..	34	60	94
Controls ..	42	52	94
Total ..	76	112	188

Professor L. S. Penrose very kindly summarized the findings shown in Table II and commented on them as follows: "Though mongols are a little more often blue-eyed than controls, the difference examined by χ^2 for one degree of freedom is 1.4—quite insignificant statistically."

It would appear, then, that anomalies of pigment metabolism, as reflected by an estimation of iris colour, are not a feature of mongolism.

I am indebted to Professor L. S. Penrose, who suggested this investigation and kindly commented on the results, and to Dr. B. H. Kirman for his interest and advice. My thanks are also due to Drs. L. Crome and J. Stern for their helpful suggestions and to the nursing staff of the Fountain Hospital for their co-operation during collection of the data.

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A Case of Ventricular Cyst Causing Hydrocephalus

In the ventricles of the brain diverticula occur as well as genuine cysts. The genuine cysts are rare and are a disease of adults rather than of children. In the literature mostly single cases are mentioned. Kelly (1951) collected 29 cases of ependymal cysts up to 1951. The youngest patient among them was 21 years old. In the Hungarian literature only Lehoczy (1929) and Kerkovits and Fehér (1954) have reported cases of cyst in the third ventricle. The case recorded here is of an unusually large cyst in the third ventricle, occurring during infancy.

CASE REPORT

At the end of 1954 a 3-months-old male infant was admitted to our department. He was the second child, and was born after a normal pregnancy and labour. Family history was of no importance. At birth the skull was normal in size, but when he was 6 weeks old it was observed to be too big for his age.

On his admission to hospital the huge cerebral skull was in striking contrast to the small facial skull and the abnormally small body. Dilated veins were visible on the skull. The anterior fontanelle was bulging and enlarged to the size of half a hand. The coronal and sagittal sutures of the skull were widely separated. The posterior fontanelle was the size of a baby's hand. The skull bones were soft. The circumference of the skull was 48 cm., that of the chest 37 cm. Signs of bilateral bronchopneumonia were present.

The infant was apathetic and his face was expressionless. The pupils were equal, dilated, and faintly responsive to light. The sucking reflex was regular. Chvostek's sign was negative. The tone of all muscles was increased, and the limbs were in extension. There were no signs of meningism. Tonic-clonic spasms in the limbs occurred several times a day. Lumbar puncture yielded only small amounts of fluid, so that fluid was drawn off from the anterior fontanelle. It was slightly yellow. Pandy's globulin test on the fluid was positive, Walter's bromide test negative. A few lymphocytes were present. The level of sugar was 71 mg. per 100 ml. and of albumin 34 mg. per 100 ml.

Bronchopneumonia persisted, and the infant became steadily worse. He died on the 48th day after admission to hospital.

Post-mortem Findings.—At necropsy the circumference of the skull in the occipito-frontal plane was 50 cm. The small fontanelle measured 8 by 5 cm.

The sutures were widely separated and the skull bones thin. The brain was very large (see Fig.), the gyri were flattened, and the substance was soft. On opening the lateral ventricles a large quantity of fluid gushed out. A cyst the size of a man's fist bulged from the third ventricle; it was full of blood, and was connected to the bottom of the ventricle by a thin peduncle. It filled the whole cavity of the ventricle, compressed the neighbouring structures, and blocked the flow of fluid. Histological examination showed that the wall of the cyst was dense fibrous connective tissue, lined inside with a single layer of flat cells. Elastic fibres were not seen. With van Gieson's stain the wall showed fuchsin red, while the inner layer of cells was orange. This layer stained red with Heidenhain's (azan) stain, the wall being aniline blue. With Foot's silver impregnation the finest fibres of the wall and the lining layer of the cells proved to be argentaffin. From these histological observations the conclusion can be drawn that the dense wall of the cyst consisted of mesenchymal tissue of embryonal character containing no elastic elements. Elements of muscle were not found. The cells lining the cyst were shown by the stains to be young endothelial cells.

We considered the case worth publishing because of the great size of the cyst and because of its occurrence during infancy.

Dissected brain. a=side ventricle; b=cerebellum; c=cyst.

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