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CONGENITAL DUODENAL OBSTRUCTION AND MONGOLISM

BY

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We have recently been impressed by the frequency with which intrinsic obstruction of the duodenum in the newborn has been associated with mongolism. During the past 25 years 32 infants with congenital atresia or severe stenosis of the duodenum have been admitted to Great Ormond Street, and of this number no fewer than 10 were recognized as mongols.

Case Reports

Case 1.—A first-born male child, three weeks premature. Mother aged 25. Admitted aged 6 weeks because of vomiting and constipation from birth. *Clinical diagnosis:* Mongolism and pyloric stenosis. *Operation:* Gastro-enterostomy for apparent "duodenal ileus." Death occurred a few hours after operation. *Necropsy:* Mongolism; stenosis of third part of duodenum, peritonitis, and organized pleurisy.

Case 2.—A male child, tenth in family, born at seven months' gestation. Admitted aged 2½ days with imperforate anus, severe vomiting, and deep jaundice with purpura. *Clinical diagnosis:* Mongolism with anal atresia. *X*-ray film suggested possible duodenal atresia. *Operation:* Relief of anal atresia. Death occurred the following day. *Necropsy:* Mongolism; atresia at junction of second and third parts of duodenum; large subarachnoid haemorrhage.

Case 3.—A full-term male child, second in family. Admitted aged 1 week with vomiting. *Clinical diagnosis:* Mongolism with duodenal atresia. Died on tenth day. *Necropsy:* Mongolism; duodenal atresia just below ampulla; partial patency of foramen ovale.

Case 4.—A male child born six weeks before term after surgical induction for hydramnios. Eighth child; mother aged 36. Admitted aged 4 days with vomiting from birth. *Clinical diagnosis:* Mongolism; prematurity and duodenal atresia. Death occurred on the sixth day. *Necropsy:* Mongolism; duodenal atresia immediately above ampulla; interventricular septal defect; cerebellum disproportionately small; undescended left testicle.

Case 5.—A male child, third in family. Admitted on day of birth with progressive jaundice. Rh incompatibility; direct Coombs's test positive. Typical mongol. *Clinical diagnosis:* Haemolytic disease of the newborn with mongolism. Death took place on second day. *Necropsy:* Mongolism; severe duodenal stenosis 1 cm. above the papilla; cerebral kernicterus; erythroblastosis foetalis; Meckel's diverticulum.

Case 6.—A first-born female child, two weeks premature; white asphyxia at birth following difficult forceps delivery. Mother aged 35. Admitted aged 3 days with vomiting from birth and passage of only mucus per rectum. *Clinical diagnosis:* Intestinal obstruction. *Operation:* Duodeno-jejunostomy for atresia at duodeno-jejunal flexure; lack of mesenteric attachment of ascending colon noted. Discharged home at 1 month of age after a stormy convalescence. Recognized as a typical mongol when seen in an out-patient clinic at 5 months of age.

Case 7.—A male child, third in family. Mother aged 36; suffered from hydramnios during pregnancy. Admitted aged 7 days because of persistent vomiting. *Clinical diagnosis:* Intestinal obstruction. *Operation:* Gastro-jejunostomy for duodenal atresia. Died shortly after operation. *Necropsy:* Mongolism; complete membranous occlusion of duodenum immediately distal to ampulla; extensive peritoneal haemorrhage; small cerebellum; undescended left testicle.

Case 8.—A female child five weeks premature, fifth of family. Mother aged 40; hydramnios during pregnancy. Admitted on third day with vomiting from birth. *Clinical diagnosis:* Atresia of duodenum. *Operation:* Duodeno-jejunostomy for atresia of proximal 4 in. (10 cm.) of jejunum. Died shortly afterwards. *Necropsy:* Mongolism; duodenal atresia immediately distal to papilla; multiple atresias of proximal jejunum; small intestine much shorter than normal; universal mesentery; cerebellum disproportionately small; partial patency of foramen ovale; failure of lobation of left lung.

Case 9.—A first-born full-term female child. Mother aged 25; hydramnios during pregnancy. Infant suffered asphyxia at birth. Admitted aged 5 days with vomiting, cyanotic attacks, and purpura. *Clinical diagnosis:* Duodenal atresia and possible mongolism. *Operation:* Peritoneal bands divided. Death on eighth day with oedema and cardiac failure. *Necropsy:* Extensive peritoneal haemorrhage; duodenal atresia immediately above the papilla; mongolism, small cerebellum; large auricular septal defect (septum secundum); pulmonary oedema.

Case 10.—A male child, eighth in family. Maternal age 39; hydramnios during pregnancy. Following severe birth asphyxia the infant began to regurgitate brown fluid. Admitted aged 5 days. *Clinical diagnosis:* Mongolism and duodenal atresia. *Operation:* Gastro-jejunostomy. Deterioration after tenth day with diarrhoea; death at 3 weeks of age. *Necropsy:* Mongolism; severe duodenal stenosis immediately proximal to the ampulla; universal mesentery; small brain; aberrant right subclavian artery; umbilical sepsis; oral thrush.

Discussion

Textbooks of surgery and paediatrics do not in general refer to this association, which in our experience has occurred with remarkable frequency, nor is it mentioned in the monographs of Benda (1947) and Engler (1949) on mongolism. In a survey of published cases of duodenal obstruction in the newborn Forshall (1947) found only five instances in which associated mongolism had been observed. More recently, however, Lanman (1949), in Boston, reported that, of 17 children with duodenal

atresia successfully treated by operation, 4 were subsequently found to be mongols, while Grove and Rasmussen (1950) found 3 mongols among 11 cases of duodenal atresia or stenosis.

One of us (L.L.R.W.) recently observed mongolism in three successive infants with duodenal atresia at necropsy, in Cardiff, and from personal communications we have learned that Miss I. Forshall, in Liverpool, has seen four cases, and Miss V. Smallpeice, at Oxford, two cases.

From our observations there would appear to be a high incidence of mongolism in infants suffering from intrinsic malformations of the duodenum, the overall frequency in 25 years being approximately one in three. It is remarkable, however, that during 1951, after our interest had been drawn to the association, all five cases of duodenal atresia or stenosis admitted to Great Ormond Street have been found to be mongols; in two of these the latter condition was not recognized until post-mortem examination. Our recent experience suggests that the frequency of associated mongolism may be even higher than one in three.

A possible explanation of the paucity of recorded instances is that most uncomplicated cases of mongolism do not come under medical observation until there is obvious retardation of mental progress, at the end of infancy or later. On the other hand, most cases of duodenal atresia or stenosis die in the neonatal period at a time when the classical appearances of mongolism, though present, are not conspicuous.

The features of mongolism in newborn infants are perhaps not as widely appreciated as they might be (Carter and MacCarthy, 1951). On clinical inspection the immediate general impression is that the baby has an odd appearance. Only a careful systematic examination will elicit the presence of some of the signs of mongolism.

The head is usually rounded, and on palpation of the sagittal suture a third fontanelle may be found. The facial contour is square; the oblique palpebral fissures are small and measure less than the interocular distance; medial epicanthic folds are usually present; the nasal bridge is depressed; the ears are poorly formed and have a crumpled appearance; the tongue tends to protrude. Fingers are somewhat short and the little finger may be incurved owing to a short middle phalanx; a single transverse palmar crease is often present; the axial triradius is in an abnormal position (Penrose, 1949). A high maternal age affords further circumstantial evidence.

At post-mortem examination the most characteristic internal finding is a disproportionately small cerebellum and brain stem; associated malformations, notably cardiac septal defects, may be encountered.

It must be emphasized that no one feature is peculiar to mongolism, nor is any sign of invariable occurrence; it is the presence of a majority of these signs, however, which enables one to make the diagnosis with reasonable certainty, even in the newborn.

In view of our observations it seems important to seek evidence of mongolism in all infants with suspected duodenal obstruction, since such finding may influence the decision to operate.

Summary

In a consecutive series of 32 infants with congenital atresia or stenosis of the duodenum a high incidence of mongolism was observed.

The frequency of this association has not been generally appreciated, presumably because most children with duodenal atresia or stenosis die during the neonatal period, when mongolism is apt to be overlooked. The features of the newborn mongol are therefore reiterated. It is felt that the early recognition of mongolism in a newborn child with suspected duodenal obstruction may be of practical importance in the management of the case.

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ACUTE INFECTIOUS LYMPHOCYTOSIS

REPORT ON A GROUP OF CASES IN A DAY NURSERY

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Acute infectious lymphocytosis was first described as a specific disease by Carl H. Smith, of New York, in 1941. It chiefly affects children below the age of 10 years, and is known to occur sporadically and also in small epidemics, particularly in institutions and among members of the same family. The literature and features of the condition were fully reviewed by Smith himself in 1947. Most of the case reports have come from North America, and the largest recorded outbreak there involved 86 persons in an institution for mentally defective and epileptic patients (Barnes *et al.*, 1949). During the last few years instances of the disease have also been reported increasingly in Europe. Thus the condition has been encountered in Finland (Klemola and Hamarinen, 1947), in Switzerland (Gsell, 1947; Landolt, 1947; Martens, 1948), in France (Debré *et al.*, 1948; Lamy, 1948; Bernard *et al.*, 1948), and in Germany (Recknagel, 1948; Koch, 1949; Heni and Zeh, 1949), as well as in Austria, Czechoslovakia, Greece, the Netherlands, Sweden, and Spain.

In this country, Kilham and Steigman (1942) first pointed out certain similarities to infectious lymphocytosis in three children whose blood showed an increase in the number of small lymphocytes and gave a negative Paul–Bunnell reaction. Later, Steigman (1946) briefly described a small outbreak affecting six children in a village in Southern England in 1942. Isolated instances of the condition in the East End of London have been reported from this hospital (Sita-Lumsden, 1947) and from Mile End Hospital (Dolphin and Popham, 1950). An account of three cases was also published in Northern Ireland (McCollum, 1949).

Six cases which appeared to originate in a day nursery in East London are described below. The first three were encountered in hospital practice.