Lesson of the Week

Do you shake hands with mothers of floppy babies?

T H H G KOH

There are very few clinical situations where the birth of a child can lead to the new diagnosis of a serious condition in the mother and her family. In the past year our unit has had two admissions which resulted in the diagnosis of myotonic dystrophy in both babies and 12 members of their families. I describe one of these two babies in whom routine handshaking with the mother led to an early diagnosis.

Case reports

The baby—A baby girl was delivered vaginally at 38 weeks' gestation after spontaneous labour. Polyhydramnios had been noted at 33 weeks. No drugs were given during labour but meconium stained liquor and type 2 decelerations were noted before delivery. At birth the baby was covered in fresh meconium with mild birth asphyxia and was resuscitated appropriately. At five minutes she was breathing spontaneously and pink in air. Initial electrolyte and blood sugar concentrations were normal. On examination she was extremely hypotonic and areflexic and had a feeble cry. She had a large cephalhaematoma, bilateral pes cavus, and mild talipes equinovarus. A differential diagnosis of perinatal asphyxia or neuromuscular disease was made. Feeding proved slow and difficult and nasogastric feeding was initially needed.

The mother—The baby was born just before midnight. The next morning the mother visited the baby and on routine handshaking showed an obvious myotonic grip. The mother was a 28 year old married Caucasian, who was unaware of her muscle disease but had noticed certain difficulties for some years: she could not easily remove pegs from the clothes line or unscrew caps from containers. She admitted to feeling "more stiff" during this and her two previous pregnancies. She thought that fetal movements had been normal during her recent pregnancy. On examination she had bilateral ptosis, percussion myotonia, mild dysarthria, inability to bury her eyelashes, and a myopathic smile. Her two daughters, aged 4 and 8, had never had symptoms or signs of muscular disease. There had been two spontaneous abortions at 5 and 7 weeks.

Examination of other members of the mother's family confirmed the presence of myotonia in her mother, her twin sister, and her two brothers. A niece aged 8 months born to the twin sister had been hypotonic since birth, although no diagnosis had been made. Examination of this baby disclosed the tent shaped mouth and considerable hypotonia. No members of the family were aware of their condition, although one of the mother's brothers, a schoolteacher in a large comprehensive, had failed to release a piece of chalk when attempting to throw it at a pupil. He had been thought by his general practitioner to have "rheumatism" because of "morning stiffness and pain in the muscles." The whole family was subsequently referred for neurological assessment and genetic advice.

Special Care Baby Unit, Hope Hospital, Salford M6 8HD
T H H G KOH, MA, MRCP, locum registrar in paediatrics

Correspondence to: Department of Paediatrics, Royal Berkshire Hospital, London Road, Reading, Berks RG1 5AN.

Discussion

In adult medicine the social and diagnostic importance of shaking hands with patients is well known but, as this case illustrated, not well practised. This sequence of events emphasises the importance of shaking hands and examining the mothers and families of floppy babies; this is especially so in the case of babies who are transferred initially without their mothers to regional neonatal intensive care units.

The detection of neonatal myotonic dystrophy may be difficult and is based on clinical suspicion. The condition is not as rare as is generally thought.[1] Since the affected babies often have respiratory problems it is common to attribute the hypotonia to perinatal asphyxia. Characteristically the clinical picture consists of extreme hypotonia, neonatal respiratory problems, joint deformities, facial diplegia with a tent shaped mouth, unexplained haematomas, and polyhydramnios.[2] Most mothers and their families are not known to have myotonic dystrophy. Thus in two series 38 out of 59 families were not known to have myotonic dystrophy at the time of delivery.[1 2] In most cases the mother has noticed the myotonia but, as the rest of the family have the symptoms, she has never thought of her disability as abnormal and therefore never reported them. Indeed, when our patient's mother was asked whether there was any family history of muscle disorder she replied "No." Early diagnosis avoids unnecessary extensive investigations for the numerous causes of hypotonia in a baby. In our case the birth of the baby led to the diagnosis of myotonic dystrophy in six members of the mother's family and to timely genetic counselling.

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References

[1] Harper PS. Congenital myotonic dystrophy in Britain I. Clinical aspects. *Arch Dis Child* 1975;**50**:505-13.
[2] Pearse RG, Howeler CJ. Neonatal form of dystrophia myotonia. *Arch Dis Child* 1979;**54**:331-8.

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Is the isolation of streptococcus viridans from a baby's sticky eye serious, and should the baby be treated?

Sticky eyes occur in about 12% of neonatal babies, but although 4% grow bacteria, with about 1/7th of this group growing streptococci, 4% of normal babies also have streptococci in the conjunctival sac. The evidence suggests that in neonates this group of organisms is usually acting as a pathogen. Treatment with topical chloramphenicol is therefore recommended.—PETER WRIGHT, consultant surgeon, London.

Pierce JM, Warden NE, Seal DV. Ophthalmia neonatorum in the 1980s: incidence, aetiology, and treatment. *Br J Ophthalmol* 1982;**66**:728-31.