collected into a sterile universal container. Skin antiseptics were not applied to the breasts and no mother was taking antibiotics. The breasts were frequently washed with soap and water. The milk samples were delivered to the laboratory and dealt with immediately. Serial dilutions of milk between 10^{-1} and 10^{-4} were made in nutrient agar (Oxoid), using a semiautomated diluter, and after overnight incubation at 37°C the bacterial species and number of colony forming units per litre were determined. Student's paired *t* test was used to measure the significance of differences in colony counts between the two milk samples.

Results

The Figure shows the bacterial colony counts in the samples of breast milk. There was no significant difference between the colony counts of the paired samples, and in only one instance did the bacterial flora of the second sample differ from that of the first.

Discussion

West and Hewitt[3] showed that 10 ml milk needs to be discarded before the bacteriological quality of the milk improves significantly. Our study shows the futility of discarding the first 2–3 ml while collecting breast milk for banking. We conclude that milk banks whose milk derives mainly from early lactating mothers should not discard the first part of the milk collected, as this will appreciably reduce the quantity of the milk without bacteriological advantage.

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Correspondence to Dr L Carroll, Department of Child Health, Leicester Royal Infirmary Maternity Hospital, Leicester LE1 5WW.

Visual deterioration as presentation of subacute sclerosing panencephalitis

HEATHER M JOHNSTON, GRAHAME A WISE, AND J GRAHAM HENRY

Prince of Wales Children's Hospital and Marsden Hospital, Sydney, Australia

SUMMARY A 7-year-old boy presented with deteriorating vision and macular degenerative changes. A month later he had developed unusual behaviour and increasing forgetfulness. An electroencephalogram showing periodic complexes, and high measles complement-fixation titres in the cerebrospinal fluid and blood, confirmed the diagnosis of subacute sclerosing panencephalitis. Four months after the onset of visual symptoms he started having myoclonic jerks.

A focal chorioretinitis has been described frequently in patients with subacute sclerosing panencephalitis (SSPE).[1] However, visual symptoms preceding the more common features of the disease—myoclonic jerks, mental deterioration, and progressive motor deficit—are rare.[2–5] We report the case of a child referred because of visual problems noted at school.

Case report

A 7-year-old boy was referred by his school to an ophthalmologist because he had been complaining for 2 months that he found it difficult to see his schoolwork. His visual acuity was assessed at 6/60 in both eyes. Bilateral macular degenerative changes were seen and thought consistent with a dystrophy.

On review one month later his mother's chief concern was an alteration in his behaviour. He had become increasingly forgetful, failing to complete simple tasks at home. The school reported him to be vague and not learning as well as previously. At this stage he was referred to the neurology clinic at this hospital.

He presented as co-operative and alert but had difficulty understanding straightforward requests during the examination. He was unable to perform simple calculations or to give his address. Physical examination was normal, apart from the eye findings. Pupils were equal and reacted to light. Fields were difficult to test because of his lack of understanding but peripherally they appeared to be intact. Visual acuity was estimated at less than 6/60 bilaterally on formal testing, but at 6/24 with pictures. On fundoscopy the discs appeared normal. The right macular area showed an outer irregular dark pigmented area surrounding a light central 'hole'.

Similar, but lesser, changes were seen on the left. A cerebromacular degeneration of the Batten type was suspected.

Among the initial investigations was an electroencephalogram which showed periodic sharp and slow wave complexes occurring every 8–10 seconds, suggesting SSPE. His cerebrospinal fluid (CSF) contained 2 mononuclear cells per mm^3 and the protein content was 0·23 g/l. IgG was increased at 0·052 g/l giving an IgG/albumin ratio of 0·45 (normal 0·04–0·24). The diagnosis of SSPE was confirmed by measles complement-fixation titres of 1 in 64 in the CSF and 1 in 2048 in the blood.

When he had been 7-months old his sister had had measles and been unwell for several days. He had had a similar rash lasting one day, but was not unwell.

Two weeks after discharge he had a generalised tonic seizure and within a few weeks was having regular myoclonic jerks. Within 6 months of diagnosis he required institutional care because of increasing motor disability and lack of responsiveness.

At this stage both discs were noted to be pale and the retinal vessels of decreased calibre. The right macular region contained pigmented and depigmented areas as before with increased light reflexes (Fig. 1). On the left some macular oedema was noted with less marked pigmentary changes than on the right but with some chorioretinitis along the vessels. An electroretinogram was normal

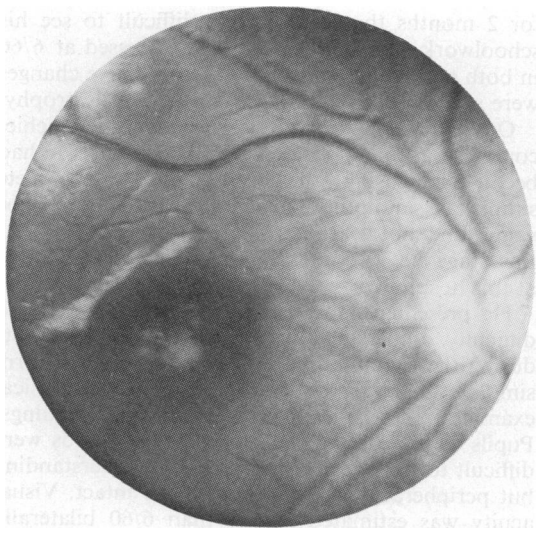

Fig. 1 *Hyperpigmentation and 'hole' formation in the right macular region with increased light reflexes lateral to this area. (Taken 9 months after onset of symptoms.)*

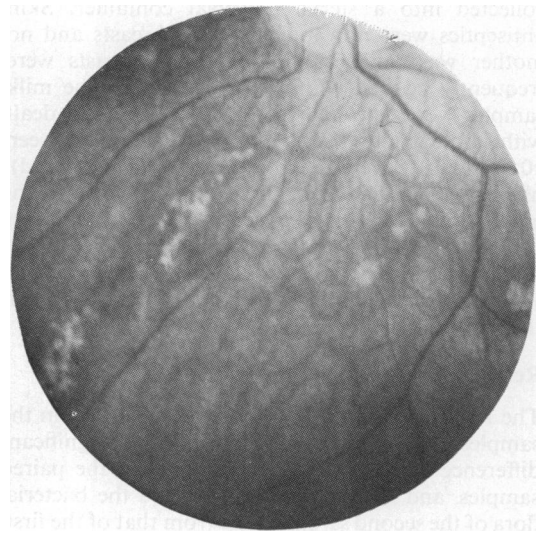

Fig. 2 *Areas of depigmentation in the lower part of the left retina. (Taken 10 months after onset of symptoms.)*

but no cortical-evoked responses could be obtained to flash stimulus. A month later further retinal changes were noted with a depigmented area extending to the periphery in the left nasal sector (Fig. 2).

Discussion

In 1970 Nelson *et al.*[3] demonstrated Cowdry type A intranuclear inclusions in ganglion and bipolar cells adjacent to an area of foveal necrosis in a child who died with SSPE. Font *et al.*[6] were able to show intranuclear inclusions in glial cells as well as in all three layers of neurons in the macular area. Electron microscopical examination showed that the inclusions consisted of filamentous structures of a paramyxovirus consistent with the measles virus.

The findings of macular pigmentary changes with 'hole' formation and focal areas of chorioretinitis should alert the clinician to the possibility of SSPE in a previously well child with visual symptoms. Early diagnosis will prevent unnecessary investigations and prolonged uncertainty for the parents.

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Correspondence to Dr Heather M Johnston, Regional Neurological Centre, Newcastle General Hospital, Westgate Road, Newcastle upon Tyne NE4 6BE.

Congenital central hypoventilation syndrome

A report of successful experience with bilateral diaphragmatic pacing

MICHAEL COLEMAN, STEPHEN J BOROS, THOMAS L HUSEBY, AND WILLIAM S BRENNOM

Department of Neonatal Medicine and Department of Surgery, Children's Hospital, St Paul, Minnesota, USA

SUMMARY Bilateral diaphragmatic pacing was successfully performed in an 18-month-old child with congenital central hypoventilation syndrome (Ondine's curse) as an alternative to long-term mechanical ventilation. Subsequent complications were related more to cor pulmonale and tracheostomy care than to the pacing itself.

Congenital central hypoventilation syndrome (CCHS), often referred to as 'Ondine's curse', is a rare disorder of ventilatory control, characterised by hypoventilation during sleep. Patients with this disorder require long-term mechanical ventilation, and most eventually develop fatal cor pulmonale. Few paediatric cases have been reported. Hunt *et al.*[1] reported the successful treatment of 3 infants with this condition, using radiofrequency diaphragmatic pacing. This report describes another successful experience using bilateral diaphragmatic pacing in the treatment of an 18-month-old child with CCHS.

Case history

A term boy, weighing 2992 g, was born after an uncomplicated pregnancy, labour, and delivery. Apgar scores were 7 and 8 at one and five minutes. During the first 8 hours of life, the infant experienced apnoeic episodes while sleeping. Serum electrolytes, glucose, calcium, and magnesium determinations were normal. Cerebrospinal fluid examination was normal. Chest *x*-rays showed no cardiomegaly or pulmonary disease. Skull *x*-rays, cranial computerised tomography, and electroencephalograms were normal. The apnoeic episodes were initially treated with nasal continuous distending airways pressure and, later, with intravenous aminophylline. Neither treatment was successful.

On the third day of life, the infant was intubated and intermittent mandatory ventilation was administered during sleep. During the next 2 months, he received therapeutic trials with atropine, caffeine citrate, progesterone, thyroxine, oral doxapram, and phenobarbitone. None of these drugs was successful in preventing hypoventilation during sleep. A tracheostomy was performed at 2 months of age. Bronchoscopy and fluoroscopy of the trachea were normal. A carbon dioxide ventilatory response during sleep showed no increase in minute ventilation, despite increases in alveolar CO_2 to 60–80 mmHg. Ventilation was normal when awake.

At 6 months of age, he was sent home with a volume preset ventilator for respiratory assistance during sleep. However, during the next 2 months, the child and his family experienced many mechanical problems with the ventilator, pulmonary toilet, and recurrent pulmonary infections. Eventually, parental stress necessitated readmission. At that time, the patient's chest *x*-ray showed mild cardiomegaly. His electrocardiogram showed right ventricular hypertrophy. Both of these findings were consistent with the diagnosis of early cor pulmonale. Cor pulmonale was treated with chronic diuretics and the administration of 30% oxygen during spontaneous wakeful breathing and during mechanical ventilation when asleep.

At 18 months of age, bilateral phrenic nerve