rence of typhoid in India and the youngest case reported was 12 days old(7).

The isolation of pneumococcus alongwith *S. typhi* in our case might have occurred as complication of trauma, as pneumococcal meningitis occurs usually as a complication of middle ear disease, with or without mastoiditis. Prematurity and trauma were the factors leading to greater susceptibility of infection in this case. The isolation of two organisms in CSF of this neonate, especially *Salmonella typhi* with *pneumococcus,* is noteworthy.

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Pendred's Syndrome

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Congenital sensori-neural deafness in association with a goitre is termed as Pendred's syndrome(1). We are reporting such a patient whose goitre was apparent at birth and the diagnosis of sensori-neural hearing loss was accomplished using auditory evoked responses during the neonatal period.

Case Report

A 22-day-old, term, male child was born to a 33-year-old mother with Grade II untreated goitre. The family consumed iodized salt at home and no other family member had goitre.

The baby had been referred with the complaint of a swelling in the neck since birth (*Fig. 1*). There was history suggestive of constipation and a hoarse cry. There was no history of any difficulty in feeding or prolonged jaundice.

On examination, the child weighed 2.5 kg with a head circumference of 34 cm.

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The baby had a low hair line and low set ears; he kept his tongue protruded and had normal skin texture. The anterior as well as the posterior fontanelle were open. The heart and respiratory rates at rest varied from 96-100 and 32-34 per minute, respectively. Systemic examination was unremarkable except for the decreased tone in all four limbs. No response was elicited to a loud sound stimulus.

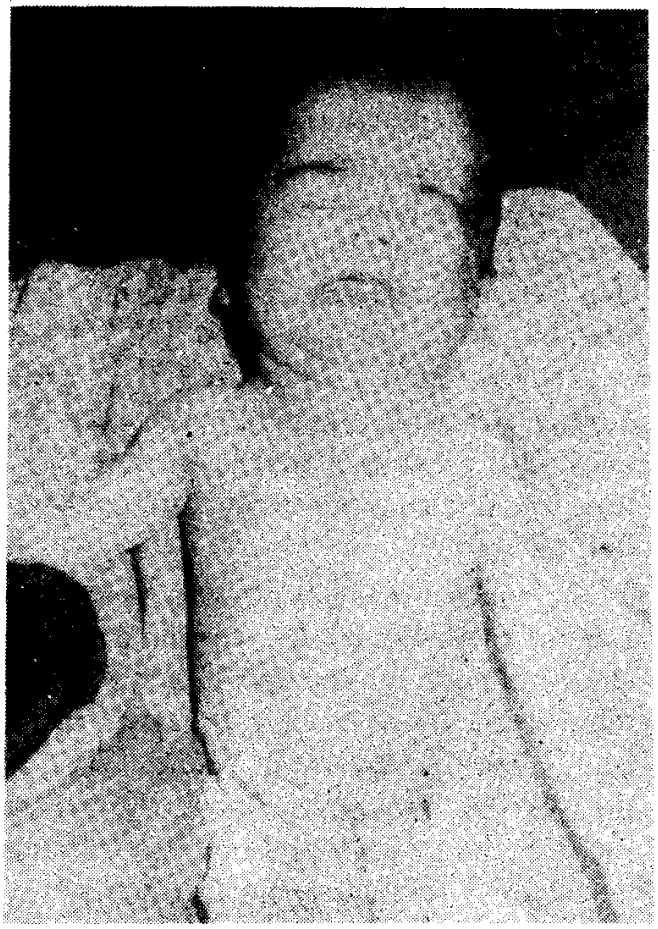

Fig. 1. The neonate with goitre.

Thyroid was enlarged (4 × 2 cm), soft in consistency and had a smooth surface. No bruit was audible. A clinical diagnosis of congenital hypothryoidism with deafness was made.

The boy's T_3, T_4 levels were 0.65 ng/ml (normal 0.8 - 1.8 ng/ml) and 6 ng/ml (normal 60-120 ng/ml), respectively. *X*-ray of the knee revealed absence of both lower femoral epiphyses. Ultrasonography of the thyroid showed diffuse enlargement without any nodules. The auditory evoked response revealed a bilateral severe sensorineural hearing loss (*Fig. 2*). Thus a diagnosis of Pendred's syndrome was confirmed. The T_3, T_4 levels in the mother were 1.24 ng/ml and 16.6 ng/ml, respectively. She was clinically euthyroid.

The child was started on 10 μg/kg of L-thyroxine daily. After 2 weeks of therapy, the child's goitre had markedly decreased in size, the cry had become normal; tone in the limbs had improved and the child was

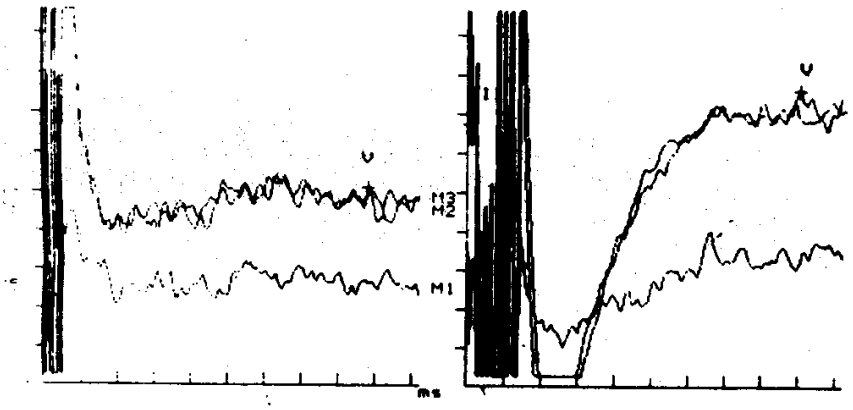

Fig. 2. Auditory evoked responses showing sensorineural hearing loss (at 80 dB).

having a normal frequency of bowel movements.

Discussion

The incidence of Pendred's syndrome has been variously reported from 1/100,000 in Sweden to 8/100,000 in England(2). No comparable figures are available for India. This syndrome has been named after Pendred who described 2 deaf-mute sisters with pronounced goitres(1). The proposed enzymatic defect was first propounded by Morgans and Trotter(3) while Fraser(4) reviewed 233 such cases from the world literature. We could find only one case report from India(5), and none diagnosed in the neonatal period.

Though the goitre formation usually commences at puberty or later, in a few cases it may be present at birth(2), as in our patient. Generally the patients are euthyroid, though hypothyroidism can exist. In a series from Denmark(6), 4 patients (out of 17) with congenital hypothyroidism had goitre developing during infancy. Earlier, surgical intervention used to be the norm for removal of the goitre(6,7) but thyroxine substitution therapy is usually effective in inhibiting the growth of the thyroid gland(2).

The hearing loss can be of all grades but is frequently very severe and most pronounced in the higher frequencies(5,7). This was the case in our patient too. An interesting description of the temporal bones in such patients have been reported. The Mondini defect, the characteristic feature of which is, while the basal turn of the cochlea is retained, the apical turn forms a large common cavity(2). This defect has been demonstrated using *X*-ray tomography(6,8). Though some workers(6,7) consider this defect as an integral part of the Pendred's syndrome, others disagree(2). It should ne noted that the character of the hearing loss with the Mondini defect does not appear to be consistent with the generally accepted theory of hearing, that the tones of the lower frequencies are registered in the apical part of the cochlea(2). Also, as a rule, the Mondini defect has been characterized as a dominantly inherited trait(7), while Pendred's syndrome has a recessive character(4), though pseudodominance can occur(2). It has not been possible to demonstrate that the sensorineural hearing loss results from the thyroid enzyme defect. It is generally accepted that Pendred's syndrome results from a pleiotropic gene, *i.e.*, a gene with 2 independent defects(2).

In recent years(2,6,9), the diagnostic evaluation of Pendred's syndrome has included a perchlorate test. The iodine metabolism in the thyroid is defective in the stage where, after having been trapped by the thyroid, inorganic iodine is incorporated in the thyroglobulin molecule where it is bound in an organic form to tyrosyl radicals. This particular defect in the thyroxine synthesis can be detected using the ability of perchlorate to release inorganic iodine from the thyroid gland. In a normally functioning thyroid gland, the organic incorporation is very rapid and only little inorganic iodine is present. Therefore, the perchlorate ions do not affect the counting rate over the thyroid after administration of radioactive iodine. We felt it was ethically unjustified to expose the neonate to radioactive iodine and hence deferred this test.

The early diagnosis of Pendred's syndrome has implications for the management of the neonate with goitre, especially the substitution of thyroid hormone (if hypothyroidism exists or to decrease the

size of the goitre). Furthermore, the quick confirmation of the sensori-neural hearing loss using auditory evoked responses has a prognostic significance and a role in genetic counselling.

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Neonatal Herpes

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'Herpes' is a Greek word meaning "to creep". Herpes infection was first described by Jean Astruc in 1736, but the occurrence of Herpes infection in the neonatal period was described by Batgnani in 1934 in a neonate with isolated keratoconjunctivitis. Neonatal herpes is by far the most serious infection attributed to herpes virus(2). We believe this to be the first case of Herpes infection in the neonate from India, successfully managed with acylovir.

Case Report

A 21-year-old primigravida delivered a live male baby by Cesarean section (Indication: fetal distress, meconium stained liquor). Birth weight was 3.4 kg. Apgar scores were 9, 10, 10. On examination, the baby had hepatosplenomegaly (liver 5 cm below the costal margin and spleen 4 cm below the costal margin) and developed respiratory distress soon after birth. He was

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