Sensorineural hearing loss in sporadic congenital hypothyroidism

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SUMMARY  Hearing acuity was assessed in 45 children with sporadic congenital hypothyroidism during adequate long-term treatment. Otoscopy was performed in each and additional tympanometry in some of them. Secretory otitis media was found in 6 and was treated medically or by inserting grommets in the eardrum. In these children, hearing acuity was assessed after the otitis had been cured. Hearing acuity was measured either by conventional monaural pure-tone audiometry (125–8000 Hz) or by binaural free field testing depending on the child's age (above and below 4 years respectively). Hearing was normal in 36 (80%) children. In the remaining 9, sensorineural hearing loss to some degree was detected affecting the higher frequencies in particular. Perceptive deafness required the use of a hearing aid in 4 children. No relationship could be found between hearing acuity and chronological age or bone age at diagnosis of congenital hypothyroidism or type of hypothyroidism. Sensorineural hearing loss is common in children with congenital hypothyroidism and should be searched for carefully and systematically to avoid difficulties related to speech and language development.

Children with congenital hypothyroidism (CH) may present various neuropsychological disturbances especially if adequate treatment is not started early in life. This has led to the introduction of screening programmes in many countries. Although it is well known that patients with CH often have articulatory speech difficulties little has been written about hearing acuity. The Pendred syndrome, a rare autosomal recessive disorder, has been studied thoroughly, and sensorineural hearing loss and goitre with or without hypothyroidism are associated. Endemic cretinism with goitre can result in severe hearing loss with deafmutism. Thus, a link between some types of hypothyroidism and hearing loss seems probable.

Recently, 3 children with CH were examined because of speech delay. Each was found to have severe sensorineural deafness requiring a hearing aid and to need auditory re-education. This prompted us to examine the hearing acuity of all our patients with CH. The aim of this study was to investigate the incidence, type, and severity of hearing loss in children with sporadic congenital hypothyroidism.

Patients

Forty-five patients including the 3 index cases were studied. There were 36 girls and 9 boys, aged between 1 10/12 years and 13 9/12 years, with both a mean and median age of 7 2/12 years (Fig. 1). Eleven patients were below age 4 years. Hypothyroidism was present in all children as shown by measurements of serum thyroxine and thyroid stimulating hormone. Five patients had been detected by neonatal screening but only one of them was diagnosed before age 1 month. The 40 other children had signs of hypothyroidism which led to the diagnosis.

The age at diagnosis of CH ranged from 14 days to 7 3/12 years with a median of 9 months. Hormonal treatment began immediately after the diagnosis of CH. All children subsequently had normal growth, bone maturation, and hormonal status. They were euthyroid at the time of auditory evaluation. Scanning of the neck, performed in 44 of them, showed an ectopic gland in 25 patients, a thyroid in situ in 7, with an aplasia of the left lobe in one. In 12 patients, no thyroid gland was detected.

Methods

Bone age was determined using the method of Sénécal et al. up to 44 weeks' gestational age and
of Greulich and Pyle for the determination of postnatal age.\textsuperscript{7} Hormonal measurements were performed by conventional radioimmunoassays and radioiodine uptake tests by routine methodology.

Neurological examination and psychometric assessments were performed according to methods described previously.\textsuperscript{2} Otoscopy was performed in all children and additional tympanometry in some. Secretory otitis media was treated medically or by inserting grommets into the eardrums, and hearing was assessed after the otitis had been cured.

Hearing evaluation was carried out by conventional pure tone audiometry (125–8000 Hz). In patients below age 4 years the thresholds were determined by the conditioned orientation reflex (binaural testing).

**Results**

Signs of secretory otitis media were found in 6 children below age 6 years. Hearing was normal in 36 (80\%) of 45 children. As shown (Fig. 2), 9 (20\%) children had sensorineural hearing losses affecting in particular the higher frequencies. In none of them had CH been diagnosed during the neonatal period.

In 5 children hearing loss was sufficiently severe to result in social disability necessitating auditory rehabilitation with the use of a hearing aid in 4 of them. In the other 4 patients hearing loss was slight causing no major social handicap.

Data for patients with mild and severe hearing losses are summarised (Table). We could not find any relationship between hearing acuity and any other variable—such as chronological age and bone age at diagnosis, aetiology of thyroid failure, duration of hypothyroidism, or neuropsychological sequelae. Of the patients with severe hearing loss 2 had an ectopic thyroid gland, in 2 no thyroid tissue could be detected, and 1 had an iodine trapping defect. Three of the 4 children with moderate hearing loss had a thyroid ectopy and 1 thyroid agenesis. IQ values were normal in 7 of the 9 children. The lowest values were found in the 5 with severe hearing losses but, as judged by the median test, the difference is not significant. We could also not find any relationship between pre, peri, and postnatal events and hearing. In the 4 children about whom the parents complained there was a delay in speech development, appropriate tests for hearing acuity were performed after considerable delay. Case 3 had a slightly malformed external ear on which surgery had been performed but hearing acuity had not been assessed at the time.

The use of a hearing aid was successful: in the 4 patients speech, psychomotor development, and behaviour improved markedly.

**Discussion**

Our data show that sensorineural hearing loss is not
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T4 = thyroxine.

Sensorineural hearing loss in sporadic congenital hypothyroidism. 421

Sporadic congenital hypothyroidism, as previously reported, is a condition in which hearing loss is not associated with any other congenital malformation or genetic abnormality. The first study to report a significant association between congenital hypothyroidism and sensorineural hearing loss was that of Rubinstein et al. (1972), who reported that hearing loss occurred in 7 children in whom hypothyroidism was diagnosed at birth. They stressed the importance of early diagnosis and treatment of congenital hypothyroidism in order to prevent hearing loss.

Sporadic congenital hypothyroidism has been estimated to occur in 0.1% of the general population. The frequency of sensorineural hearing loss in children with sporadic congenital hypothyroidism is 33%. These findings support the hypothesis that sensorineural hearing loss is a common complication of congenital hypothyroidism.

In the study by Rubinstein et al., the authors found that sensorineural hearing loss was present in 33% of the children with congenital hypothyroidism. This high prevalence suggests that there may be a genetic or environmental factor that predisposes these children to the development of sensorineural hearing loss.

The relationship between congenital hypothyroidism and sensorineural hearing loss is not well understood. Further research is needed to clarify this association and to determine if there is a genetic component.

It is important to note that the presence of sensorineural hearing loss in children with congenital hypothyroidism does not necessarily mean that the hearing loss is due to hypothyroidism. Other factors, such as prematurity or congenital infections, may also contribute to the development of hearing loss.

Despite these limitations, the findings of Rubinstein et al. highlight the importance of early diagnosis and treatment of congenital hypothyroidism in order to prevent hearing loss. It is recommended that children with congenital hypothyroidism be screened for hearing loss early in life, in order to identify those at risk and provide appropriate interventions.

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congenital hypothyroidism. A sensorineural hearing loss to a different degree was detected in 9 children, in 4 of whom deafness was so pronounced as to require auditory rehabilitation. This suggests that the frequency of deafness in CH may be about 10%—that is about one hundred times higher than in a control population in whom it occurs in roughly one per thousand births.\(^{17}\) Unlike Crifo et al.,\(^{16}\) we found hearing loss in some patients with an ectopic thyroid gland.

Secrectory otitis media was diagnosed in 6 children all aged below 6 years. These findings suggest that children treated for CH do not suffer from an increased susceptibility for middle ear effusion.\(^{18}\)\(^{19}\)

It is not known whether hearing loss is progressive. A longitudinal study is required to answer this important question.

The reason for sensorineural hearing loss in patients with CH is unknown although it seems likely that it is caused by an intrauterine abnormality. This fits in with the observation in hypothyroid animals of immature development of the organ of Corti in the cochlea. In fact, in animals rendered hypothyroid with propylthiouracil or radioactive iodine before or immediately after birth there are severe morphological and ultrastructural abnormalities in the inner ear structures—such as immaturity of the hair cells and cells of the inner spiral sulcus, with distortion of the tectorial membrane.\(^ {20-24}\) Sensory and supporting cells present immature characteristics with abnormal persistence of kinocilium.

Whatever the explanation may be, the clinician should look for hearing loss in any patient with CH. In such children, hearing acuity should be assessed systematically and carefully. Indeed, hearing loss may contribute to the many factors that impede normal psychomotor development, especially speech understanding and language development. Perceptive hearing loss should be diagnosed early and be properly treated.

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References


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