

## 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 monoaural 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.<sup>1,2</sup> This has led to the introduction of screening programmes in many countries.<sup>3</sup> 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,<sup>4,5</sup> 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 S n cal *et al.* up to 44 weeks' gestational age<sup>6</sup> and

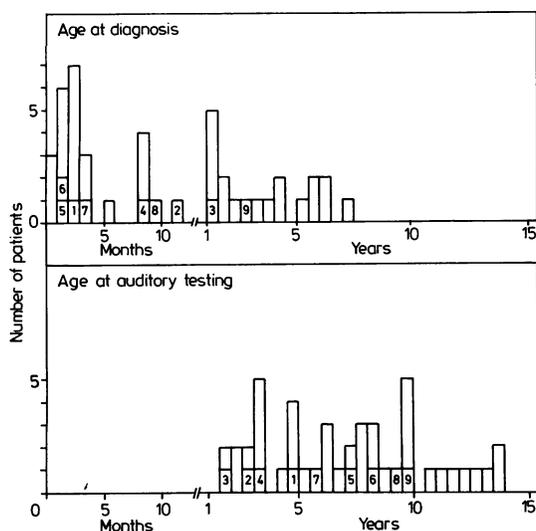


Fig. 1 Number of patients studied in relation to age of diagnosis of congenital hypothyroidism and to age at auditory testing. The numbers in the rectangles refer to the patients with sensorineural hearing loss summarised in Table.

of Greulich and Pyle for the determination of postnatal age.<sup>7</sup> 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.<sup>2</sup> 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

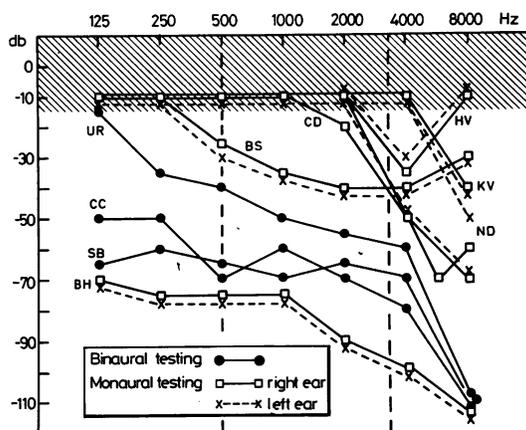


Fig. 2 Audiograms of the 9 patients with sensorineural hearing loss. The shaded area represents the results obtained in a normal population; the frequency area that is important for the perception of speech lies between the broken lines.

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

Table Patients with congenital hypothyroidism and sensorineural hearing loss

Case	Sex	Thyroid scan	Bone age at diagnosis of CH	Age at start of treatment (years)	T4 (µg/100ml)	Age at detection of hearing loss (years)	Neuropsychological defects	IQ	Complaints	Hearing aid
<b>With severe hearing loss</b>										
1	M	Absent	Prenatal	0 3/12	<0.5	4 6/12	++	52	No speech	+
2	F	Ectopic	3 months	1 0/12	1.4	2 10/12	++	98	No speech	+
3	F	Ectopic	6 months	1 2/12	<1.0	1 10/12	++	70	Speech delay	+
4	F	Trapping defect	1-3 months	0 8/12	0.4	3 3/12	++	100	Speech delay	+
5	F	Absent	Prenatal	0 2/12	0.4	7 4/12	++	106	Speech delay	-
<b>With moderate hearing loss</b>										
6	F	Absent	Birth	0 2/12	0.5	8 5/12	+	113	None	-
7	F	Ectopic	Prenatal	0 4/12	1.2	5 7/12	+	98	Speech delay	-
8	F	Ectopic	1-3 months	0 10/12	0.9	9 2/12	+	96	Speech slow	-
9	F	Ectopic	9 months	2 6/12	1.8	9 7/12	+	112	Speech difficulties	-

T4 = thyroxine.

rare in patients with CH. It is worth remembering that, as previously reported, articulatory speech defect is fairly common in congenital hypothyroidism, as it is also in some children with hypothyroidism of later onset.<sup>2</sup> It is tempting to wonder whether some patients have an undiagnosed hearing loss.

Although in this study the number of children diagnosed by neonatal screening is too small to draw any firm conclusion, none of them was found to have a hearing loss.

The frequent occurrence of perceptive deafness in patients with CH is surprising. In fact, despite excellent data obtained from experiments on animals showing the importance of thyroxine in the development of the organ of Corti, few clinical data are available. Clinical studies have mainly been concerned with two specific topics, endemic cretinism and Pendred syndrome.<sup>8-10</sup> Endemic cretinism results in endemic hypothyroidism with compensatory goitre, growth retardation, and deafmutism. In Pendred syndrome there is an association of sensorineural hearing loss with dysmorphogenesis in which oxidation and incorporation of iodine into thyroglobulin are deficient. This results in the development of goitre with a positive perchlorate test. However, neither of these conditions is present in most children with sporadic CH.

Hearing acuity has not been studied systematically in children with sporadic congenital hypothyroidism. We found only a few reports on this subject, and except for one, all were in otological journals.<sup>11-16</sup> The reports relate almost exclusively to adults with few children among the patients studied.<sup>13 14 16</sup> Moreover, the patients had various types of hypothyroidism—congenital and acquired. The data from these papers do not allow one to draw clear conclusions about the relationship between CH and hearing loss on the one hand and the duration and degree of hypothyroidism on the other. Even the precise number of children investigated is not defined. Moreover, in the study of Crifò *et al.*,<sup>16</sup> 33% of the patients were inadequately treated. This might further have influenced the audiological findings since we know from Rubenstein *et al.*<sup>13</sup> that sensorineural hearing loss may be aggravated by inadequate treatment.

Rubenstein *et al.*<sup>13</sup> mentioned that hearing loss occurred in 7 children in whom hypothyroidism was diagnosed before age 6 months. They stressed the importance of early diagnosis and adequate treatment and concluded that 'hearing dysfunction is a part of the total clinical picture of congenital sporadic hypothyroidism'.

Therefore the present study is the first on hearing acuity in children adequately treated for sporadic

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.<sup>17</sup> Unlike Crifò *et al.*<sup>16</sup> we found hearing loss in some patients with an ectopic thyroid gland.

Secretory 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.<sup>18 19</sup>

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.<sup>20–24</sup> 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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