CONGENITAL HEARING LOSS IN MALTA
A SURVEY

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The congenitally deaf infant who acquires deafness prior to development of language present special problems when compared to other hearing impaired indivudals. This is because both the development of speech and of language depend on adequate hearing. As speech and language are our prime means of communication, the congenitally deaf child is automatically also handicapped in his psychological development, social adaptation and work adaptability.

A deaf child's environment is limited to visual and tactile stimulation. He misses the comforting sounds of his mother;'s voice, the naunces of speech. He misunderstands and his reactions are often misunderstood. This might result in emotional and behavioural problems. In short a deaf child is faced by a multiplicity of problems.

Hearing loss is not an obvious handicap and is often missed because it is masked by the emotional and other problems complicating it. Early diagnosis of hearing impairment is critical to enable efforts to be made to help this child make full use of what hearing he has. The totally deaf child will develop no speech or language unless expertly helped.

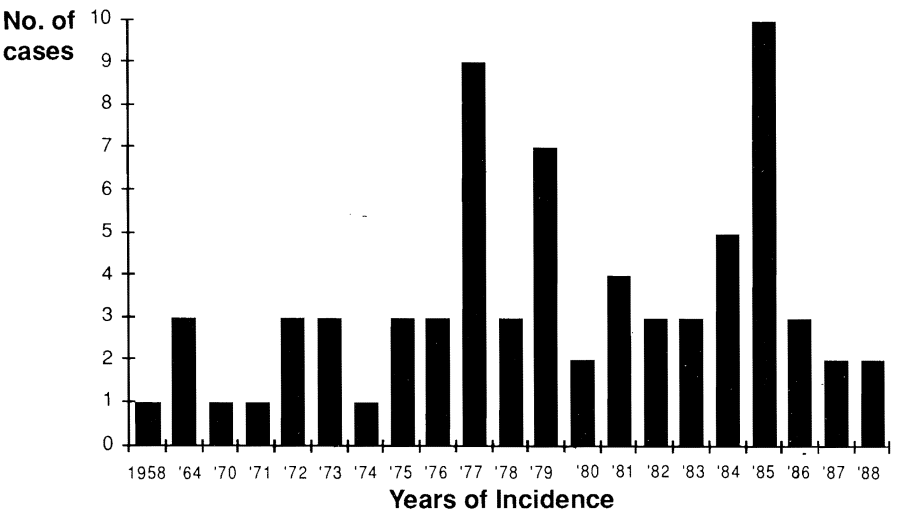
The partially deaf child will show retardation of speech and of language development, which will remain faulty.

The average incidence of congenital hearing loss in European countries is between 2 to 3 per 1,000 births, a very small number of cases, which renders identification and early diagnosis even more difficult unless a special effort is made in this direction. With these facts in mind and as part of a projected wider study, we reviewed cases of congenital hearing loss presenting on a 20 year period.

Our main aims were:
1) to establish the incidence of congenital hearing impairment in the Maltese islands;
2) to determine the common causes;
3) to highlight any difficulties in early diagnosis and management of these children;

1. INCIDENCE

TABLE 1



A total of 64 cases were collected over a period between 1970- 1989. Another 4 cases were added although diagnosed in 1958 and 1964 respectively.

This is probably not a complete figure for a number of reasons mainly because of people seeking treatment elsewhere, emigration, etc., but classification of these cases on an annual basis would appear to show an average of 3 new cases diagnosed per year with the exception of the years 1977, 1979 and 1985. The increase over these years coincided with rubella epidemics.

2. AETIOLOGY

All cases were classified as shown in Table II. The major cause of prenatal hearing loss would appear to be viral diseases with rubella responsible in 25 cases (33%). There was one case of cytomegalovirus infection, one each of measles and influenza and 2 of 'pyrexia of unknown origin.' This category should therefore all but disappear following the national immunisation campaign against Rubella.

In four patients there was a history of pre-eclamptic toxaemia, in 2 threatened

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abortion and there was one case of a diabetic mother giving birth to a deaf child.

There was a strong family history in 14 of the cases in the form of a specific hearing loss, poor speech and language development, and/or delay in development of speech. Of these, 2 cases were classified as specific syndromes.

The use of drugs in pregnancy as a possible cause of hearing loss was limited to 4 cases. The drugs involved were salicylates in 2 cases, non-steroidal anti-inflammatory drugs in one case and anti-histamines in another.

One woman was exposed to X-rays at four months gestation and another underwent surgery under general anaesthesia at 12 weeks p[pregnancy. Twenty four cases (30%) could be classified as premature birth or of low birth weight, this being 2 to 3 kg or less.

Twenty two cases suffered some form of neonatal jaundice the severity of which could not be determined. Two cases required exchange transfusion, one of which was secondary to Rhesus incompatibility. Birth trauma with anoxia accounted for 10 cases (13%).

Taking into consideration the annual birth rate, it would appear that management during pregnancy and delivery account for a minuscule percentage of causes of hearing loss.

The post-natal group have 3 cases secondary to the exanthemata (scarlet fever, measles and mumps encephalitis); one followed routine vaccination; 2 febrile convulsions the cause of which is unknown and their significance doubtful; 3 on meningitis, 2 of whom also received ototoxic drugs (gentamycin). Two cases followed severe head injury.

Only 4 cases fell into the "idiopathic" group, i.e. about 8%. This would appear to be considerably lower than expected by other authors (1,2) and this possibly because of our smaller numbers. It is moreover envisaged that with better understanding of genetic causes this group will fall still further.

TABLE II CAUSES OF HEARING LOSS

I. PRENATAL		
A. Genetic	1. Strong Family History	12
	2. Specific Syndrome	2
B. Non Genetic	1. Diseases during Pregnancy	
	a. Rubella	25
	b. Other Viruses	1 measles
		2 influenza
	c. Others	2 P.U.O.
	B. Toxaemia	5 cases
	C. Maternal diabetes	1 case
	D. Nephritis	Nil
	E. Threatened Abortion	2 cases
	II. Drugs During Pregnancy (2 salicylates, 1 Non Steroidal antiflammatory, 1 antihistamine)	4 cases
	III. Other 1 had general anaesthesia during pregnancy; 1 underwent X-Ray Examination	2 cases:
II. PERI-NATAL		
	1. Prematurity	24 cases
	2. Haemolytic disease of the new born (2 required exchange transfusion).	22 cases
	3. Birth trauma - anoxia	0 cases
III. POSTNATAL		
	A. Genetic possibly 1 case	
	B. Non-genetic	
	a. Infectious diseases: 1 scarlet fever; 1 measles; 1 mumps encephalitis; 1 post vaccination; 3 meningitis; 3 febrile convulsion	
	b. Trauma: 2 cases of head injury	
	c. Ototoxic drugs: 2 cases (gentamycin)	
IV IDIOPATHIC	4 Cases	

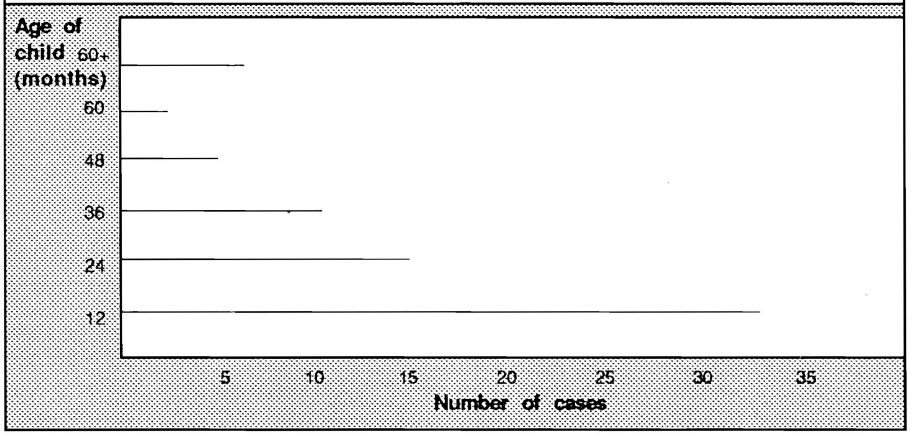
3. AGE AT DIAGNOSIS

It has long been established that early diagnosis and management of deafness is of the utmost importance (3). The ideal should be not later than 18 months.

Diagnosing early deafness is not simple and as a recent EEC study (4) showed 90% of cases had not been diagnosed by the first birthday, and as many as 50% were still not diagnosed by the age of three.

50% of our cases were diagnosed in or around the 1st year of life, 25% between 2 and 3 years and 25% beyond the 3rd year.(Table 3). It would appear at first hand therefore, that we are not doing too badly at the early diagnosis of this handicap. This could be explained by the centralisation of medical facilities at our Hospital and by adequate awareness of the problem in both the medical and teaching professions. Closer examination, however, does not show any place for complacency in this respect. Of the 68 cases considered, only 4 fell in the idiopathic group. Sixty four cases were in the so-called 'at risk' category and with the proper co-ordination of services should have been picked up at an even earlier stage. The case of Waardenberg Syndrome was only diagnosed at the age of 4 years when she at first attended school; one child with cerebral palsy at 3 1/2 years; one with monoplegia and epileptic seizures at 3 years; another who required an exchange transfusion for Rh incompatibility wa diagnosed at 6 years of age. The great majority of cases were self-referrals usually because the parents' attention was drawn to the child's apparent apathy and non-response to sound and because of poor speech development. In many cases this hearing loss was overshadowed by more obvious handicaps and missed for years though the child was under medical supervision.

TABLE III AGE AT DIAGNOSIS



4. MANAGEMENT

We were mostly concerned with:-
a) The age at which the child was fitted with a hearing aid;
b) The time interval between diagnosis and supply of the aid;
c) What other help, if any, was given in the way of speech therapy by peripatetic teachers, use of apparatus etc.

Table IV shows the age at which the child was fitted with a hearing aid. Of the 33 patients diagnosed in the first year of life, only 8 had been fitted with a hearing aid at the age of one year. Another 7 had been fitted by the age of 18 months and 5 within 2 years. More than half of our patients were therefore supplied with a hearing aid at too late a stage in their development for them to obtain maximum benefit. This is not to be taken as showing the service offered in a bad light. Many of these cases were diagnosed late, while others fell victims to parents' mistaken and misguided decisions. The normal reaction of these parents ranged from initial shock, denial, anger (often at the clinician and the team trying to help) and ultimate acceptance - a time consuming process, during which the child languishes without his hearing aid in a silent world and during which his language and emotional development remain stunted.

Around 50% of patients received their hearing aids within 6 months of diagnosis, 15% within 12 months, another 13% within 18 months and another 15% within 2 years or more. (Table 5) Again the reasons mentioned above may be playing a part. It does seem a pity however that 20 patients diagnosed before the age of 2 years all ended up being fitted with a hearing aid well after the age of 2 years.

When one considers the importance of introducing sound to the child's consciousness as early as possible in life if he is to stand any chance in developing speech and language one can understand that one month (let alone six) is too long.

Out of a total of 68 children, 19 received speech therapy while 49 did not (Table 6). Out of 19 who received speech therapy, 8 suffered from moderate to severe hearing loss and could expect some benefit from speech therapy while the other 11 suffered from severe to profound hearing loss and were correspondingly less likely to benefit from simple speech therapy. Of the 49 children not receiving speech therapy, one suffered from mild hearing loss and probably required no therapy, while 13 suffered from moderate to severe hearing loss missed out on this form of help.

It would appear that referral for speech therapy was haphazard and often the initiative to contact the speech therapist was taken by the parents themselves. In contrast the great majority enjoyed the help of a peripatetic teacher in varying periods of one to two hours per week. None of the children attending for speech therapy wore special aids.

5. EARLY DIAGNOSIS

The problem of congenital hearing loss is not a large one in numerical terms even when contrasted with the more widespread problem of acquired hearing loss in the general population. It consists basically in isolating 3 deaf individuals from an annual group of 5,000 live births. Various methods have been tried but none are fool-proof.

i) Screening Programmes: are available in many European countries. The aim is to identify the hearing impaired some time in the first year. This is supplemented by pre-school screening in an attempt to further 'close the net'. Results have not lived up to expectations and an EEC study (5) showed only 55% of children with a loss of 50 dB or more were identified before the age of 3.

In a hospital-based obstetric service such as ours, neo-natal screening is a possibility provided an effective method is available. There are problems however:-
a) It has been estimated that screening on neo-nates using the auditory response cradle in an area with a live birth rate of 3,000 annually required a full time staff of three at an estimated cost of Lm3,000 to Lm6,000 per case (6). Each case

TABLE IV AGE OF FITTING OF HEARING AID

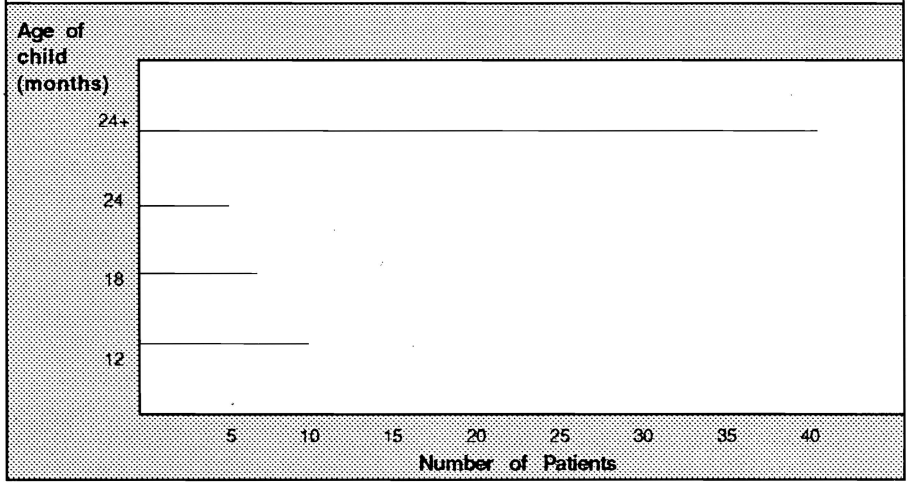
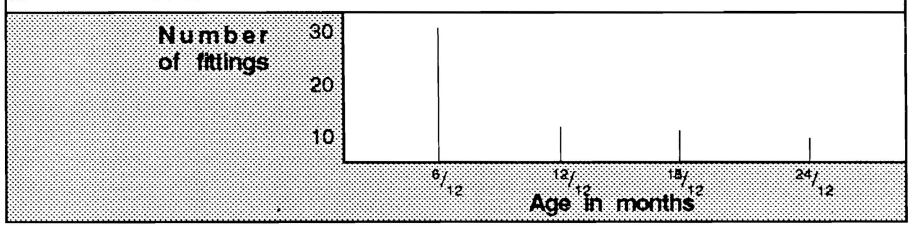


TABLE V INTERVAL BETWEEN DIAGNOSIS AND FITTING OF HEARING AID



identified would then have to be further investigated by electrophysiological means, behavioural studies etc. What is just as important is the availability for the rehabilitation of these infants. Fitting of hearing aids in neo-nates requires even more skill and experience than the fitting of an older child. Unless these facilities are available, neo-natal screening doe not appear to be a viable proposition.

b) The "at risk" register - Many (7 and 8), have expressed doubts as to the usefulness of such registers which are generally accepted as being incomplete. However in a small country like ours and especially with its now mainly hospital-based obstetric services, this method could be advantageous. From the survey carried out it appears that 95% of cases would have been immediately identified had an 'at risk' register existed. This would of course require the co-operation of the Paediatric and Obstetric Departments but that should not prove too difficult to obtain.

c) Education:- Educating the general public and parents in particular about the possibility of deafness in their offspring should prove helpful and especially useful in a small country like Malta. More than 90% of the cases in our survey were self-referred on the basis of parental suspicion of hearing loss.

6. REHABILITATION

Rehabilitation of a congenitally deaf child is a complex problem involving not only the child and his parents but also their progressive relations with a group of professionals together with the interactions between these groups of people. The present situation is not exactly an ideal one. The child and his parents find themselves shuttled between one department and another where different people offer differing or even conflicting opinions and advice. The only beacons of stability for these people in these Islands have been the lay associations for the deaf which have provided sterling services on an emotional and information level.

The recent introduction of 'Case Conferences' is a possible step in the right direction but there is no doubt that further development of the Audiology Unit at St Luke's Hospital together with more centralisation and greater participation of the Department of Speech Therapy working hand in hand with the Department of Paediatrics and Neonatology is the answer.

The involvement of the Department of Education as this stage would involve home visits by peripatetic teachers but the process of rehabilitation should be soundly established by the time the child reached school age.

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TABLE VI CHILDREN RECEIVING SPEECH THERAPY IN RELATION TO SEVERITY OF HEARING LOSS

Receiving Speech Therapy	8 cases Moderate to Severe hearing loss 11 cases Severe to Profound hearing loss
Not Receiving Speech Therapy	13 cases Moderate to severe hearing loss 36 cases Severe to Profound hearing loss