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Type II Waardenburg Syndrome

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Waardenburg in 1951 described a syndrome of lateral displacement of the medial canthi, partial albinism and deafness. Hageman *et al.*(1) in 1977 reviewed all the cases reported and classified them into three types: Type I, with dystopia canthorum (99% of all cases); Type II, without dystopia; and Type III, pseudo Waardenburg. Type III is described only in 2 patients of one family and has not yet been accepted as a distinct entity(2).

The first Indian report of Waardenburg syndrome appeared in 1962(3) and till 1978, 9 cases were reported. All of these were of Type I. Six cases of a possible variant of Waardenburg syndrome without dystopia but with Hirschsprung disease were described in 1981(4). We report here 2 cases of Type II Waardenburg syndrome.

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Case Reports

Case I was 28 years old when he reported to us for deafness. Born of unrelated parents he has 5 siblings all of whom are alive and normal. He has been deaf and dumb since birth. On examination he was of normal built and features. He had no preauricular sinus, white forelock or dystopia canthorum. His irises were bilaterally hypopigmented, blue and fundus was normal. Audiometry revealed bilateral profound sensorineural hearing loss which was partially helped by a hearing aid.

Case II is the $3\frac{1}{2}$-year-old daughter of Case I from a nonconsanguinous marriage. Her only sibling is normal. This child also has normal features and growth. She has no white forelock, hypopigmented patches of the skin or dystopia canthorum. Her iris were also bilaterally hypopigmented (*Fig.*). The audiogram revealed bilateral profound sensorineural deafness which was partially helped by hearing aids.

Discussion

Waardenburg described this syndrome of partial or complete heterochromia irides (25% expressivity) congenital deafness and dumbness, sometimes partial or unilateral (20%), white forelock (17%), lateral displacement of the inner canthi and lacrimal puncta (99%), hyperplasia of the base of the nose (78%), and hyper-

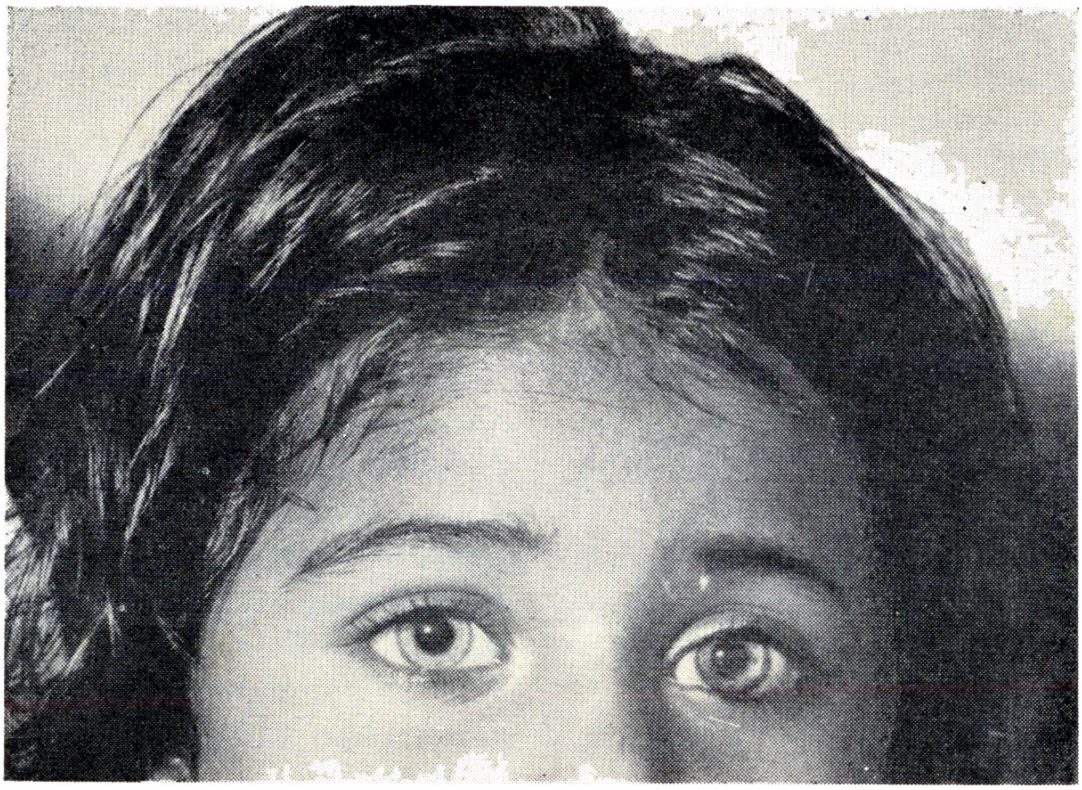

Fig. Case II showing light coloured iris.

plasia of the central section of the eyebrows (45%), due to an irregularly dominant gene, the expression being variable with a frequency of 1 : 40000(5).

The most disabling part of the syndrome is congenital deafness and it was the presenting complaint of the 2 cases that came to us. Congenital deafness defect appears to be in the organ of corti, with atrophic changes in the spiral ganglion and nerve and sparse, poorly myelinated fibers in the cochlear root of the 8th nerve(6). Both our cases had heterochromic irides. However, neither had a white forelock or cutaneous hypopigmentation.

Lateral displacement of the inner canthi is the most penetrating of the expressions of the syndrome and is seen in 99% of the cases. In this, the distance between the medial canthi is increased but the interpupillary distance and distance between the lateral canthi remain normal. This differentiates it from hyperteleorism. Waardenburg attributes it to hyperplasia of the medial frontal process and portion of the two medial and lateral processes during embryonic development. This feature was not found in the 2 cases we saw. This rare variety of Waardenburg syndrome is classified as Type II by Hageman *et al.*(1) Deafness appears 1 : 4 in Type II disease, while it appears only 1 : 8 in Type I and this is a point of relevance to genetic counselling.

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Meckel Gruber Syndrome

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Meckel Gruber syndrome, also known as dysencephalia splanchnocystica, is characterised by encephalocele, polydactyly and polycystic kidneys(1). Other commonly associated abnormalities are prenatal growth deficiency, microcephaly, micrognathia, cryptorchidism and clubfeet. The disorder is inherited by autosomal recessive transmission.

The syndrome was originally described by Meckel(1) in 1822, later by Gruber(1) and more recently brought to recognition by Opitz *et al.*(2). More than 50 cases of this severe disorder have been reported. Verma *et al.*(3), have reported a case of Meckel Syndrome from India. However, the reported case did not have some of the important features of the syndrome. Moreover the diagnosis of polycystic kidneys could not be confirmed by an autopsy. We report here a case in which, apart from the most commonly found manifestations, some uncommon and unique features and some as yet unreported manifestations were found.

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Case Report

A 40-year-old woman of Charan community (Hindu) was admitted to Irwin Group of Hospital, Jamnagar with history of 9 months pregnancy and absent fetal movements. She was para 6, gravida 6. She and her husband were consanguinous, husband being the paternal cousin of first generation of his wife.

On examination intrauterine death was suspected and labour was induced. A still-born male baby was born by vertex presentation, 5 siblings of the baby were normal and there was no family history of similar disorder in the past.

On examination the baby's weight was 2.0 kg, height 35 cm, upper segment 23 cm, lower segment 12 cm, span 32 cm, head circumference 27 cm and chest circumference 30 cm. There was microcephaly with occipital encephalocele and sloping forehead, cleft palate, lobulated tongue, tongue tie and 16 neonatal teeth on upper and lower jaws each (*Fig.*). Baby had short neck, absent nipples and limb anomalies. Right hand had postaxial polysyndaetyly, *i.e.*, 6 fingers with fusion of last two. Left hand had postaxial polydactyly. Both feet also had 6 toes. There was bilateral talepes equinovarus. Baby also had arthrogryposis in the form of contractures