Case Report

BIOMEDICAL Journal of Scientific & Technical Research

ISSN: 2574 -1241

DOI: 10.26717/BJSTR.2022.45.007181

Association of Crouzon Syndrome with Meniere's Disease

Sandeep Aggarwal[1]*, Shallu Aggarwal[2] and Manmeet Kaur Sodhi[3]

[1]*Pediatrics, Working as Assistant Professor, Department of Pediatrics, Government Medical College, India*

[2]*Ophthalmologist, Working as eye surgeon Head of Department, Department of Ophthalmology, District Hospital, India*

[3]*Department of Pediatrics, Government Medical College, India*

***Corresponding author:** Sandeep Aggarwal, Working as Assistant Professor, Department of Pediatrics, Government Medical College, Amritsar, India

ARTICLE INFO

Received: July 13, 2022

Published: July 25, 2022

ABSTRACT

Citation: Sandeep Aggarwal, Shallu Aggarwal, Manmeet Kaur Sodhi. Association of Crouzon Syndrome with Meniere's Disease. Biomed J Sci & Tech Res 45(2)-2022. BJSTR. MS.ID.007181.

Objective

To create awareness regarding association of Crouzon Syndrome with Meniere's disease

Introduction

Crouzon syndrome affects the first branchial arch, which is the precursor of the maxilla and mandible. This is an autosomal dominant birth defects that results in abnormal fusion between bones in the skull and face, resulting in an abnormally shaped head, face, and teeth. It is the most common type of syndromic craniosynostosis. Crouzon syndrome shares many of the same features as Apert syndrome [1]. Meniere's disease (MD) is a disorder of the inner ear that causes vertigo attacks, fluctuating hearing loss, tinnitus and aural fullness [2].

Case Report

A 7-year-old boy reported with chief complaint of frequent fall, vertigo and nausea. Earlier that child was under treatment of ophthalmologist for amblyopia, strabismus and refractive errors. She referred case to paediatrics department (Figure 1). Child's appearance was not normal and there was short and broad head, exophthalmos (bulging eyes due to shallow eye sockets after early fusion of surrounding bones), hypertelorism and psittichorhina (beak-like nose) and mild to moderate mental retardation. Abnormal shape of the head was noted by the mother ever since he was 6 months, and the severity has gradually increased. There was history of poor school performance (Figure 2). Mother said that many times he was not listening or responding to call. Later child told us about abnormal sound (beep like) in ear when specifically asked by leading question. A diagnosis of Crouzon Syndrome with Meniere's disease was established (Figure 3).

Discussion

In Crouzon syndrome skull and facial bones, while in development, fuse early or are unable to expand. Thus, normal bone growth cannot occur. The phenotypic features of Crouzon's syndrome may be absent at birth and may evolve gradually during the first few years of life [3,4]. It is commonly inherited as an autosomal dominant trait, with complete penetrance and a variable expressivity, but about one third of the cases do arise spontaneously. The male to female preponderance is 3:1[5]. With the advent of molecular technologies, the gene for Crouzon's syndrome

could be localized to the FGFR2 gene, at the chromosomal locus 10q25.3-q26, and more than 30 different mutations within the gene have been documented in separate families [6]. The appearance of an infant with Crouzon's syndrome can vary in severity from a mild presentation with subtle midface deficiency to severe forms with multiple cranial sutures fused and marked mid face and eye problems. Low-set ears are a typical characteristic. During normal development, the ears "travel" upward on the head; however, in Crouzon patients, this pattern of development is disrupted. Ear canal malformations are extremely common, generally resulting in some hearing loss. Patients sometimes exhibit malformations of the external ear and/or the middle ear, such as malalignment of the pinna, conductive hearing loss caused by middle ear effusion (or fluid in the middle ear) and perforation to ossicular fixation, intratympanic bony masses and ossicular anomalies. In particularly severe cases, Ménière's disease (light-headedness, vertigo, or ringing in the ears) may occur.

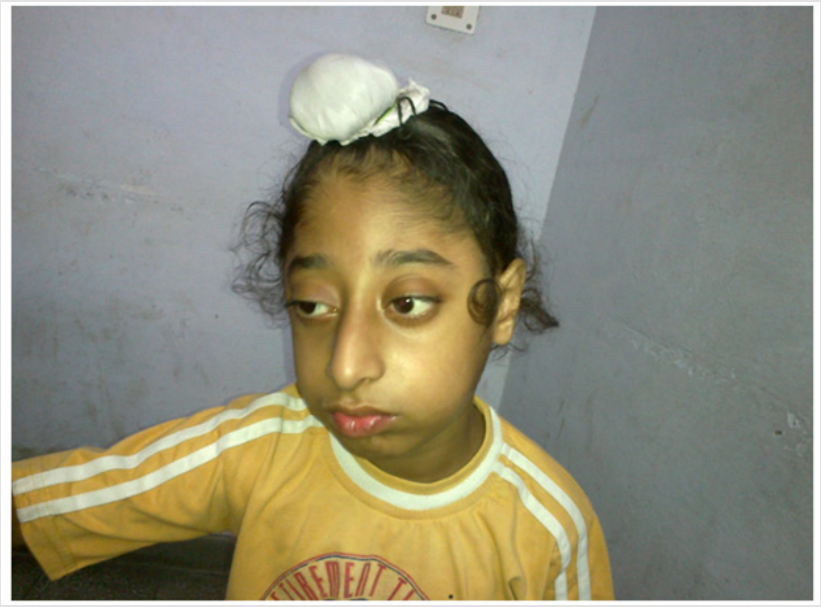

Figure 1.

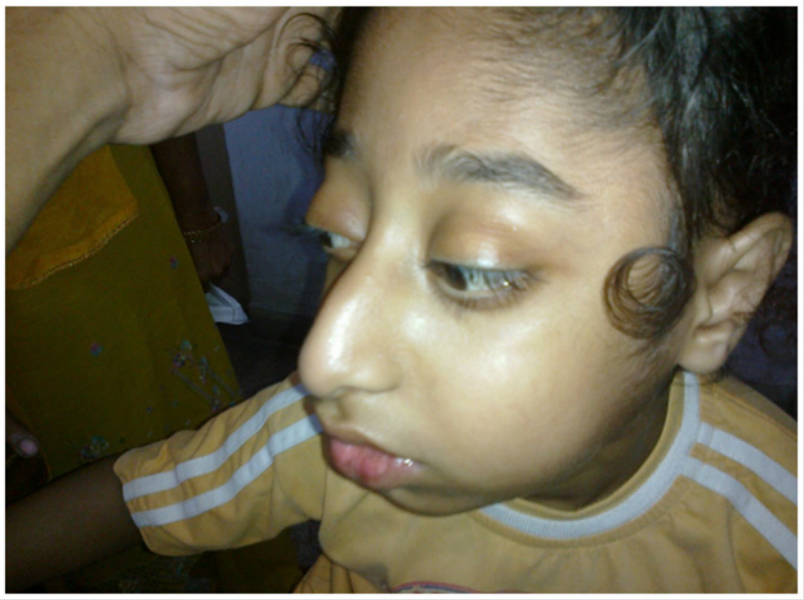

Figure 2.

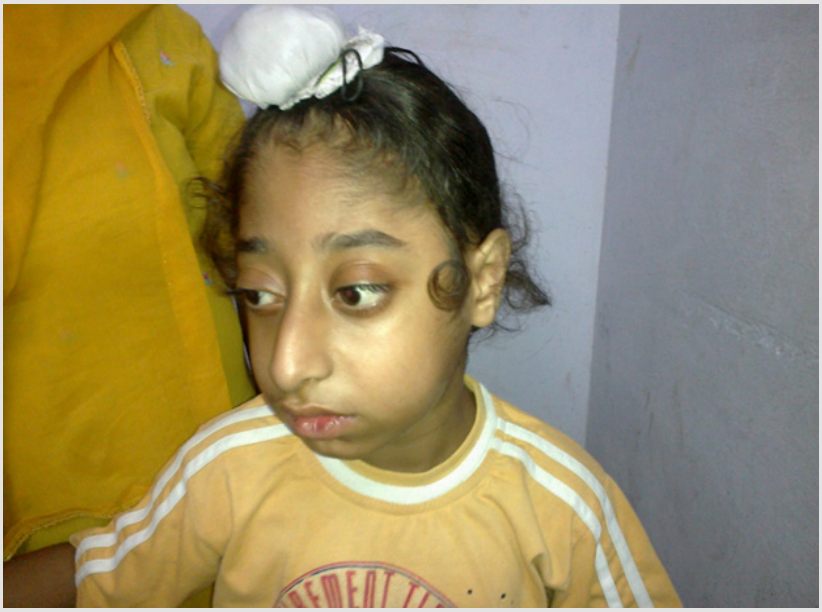

Figure 3.

Conclusion

Most patients with craniosynostosis have recurrent otitis media with effusion, causing episodes of conductive hearing loss throughout their lives. we recommend routine visits to the otolaryngologist to screen for otitis media with effusion throughout life.

References

1. Carinci F, Pezzetti F, Locci P, Becchetti E, Carls F, et al. (2005) Apert and Crouzon syndromes: clinical findings, genes and extracellular matrix. J Craniofac Surg 16(3): 361-368.
2. Nakashima T, Pyykkö I, Arroll MA, Casselbrant ML, Foster CA, et al. (2016) Meniere's disease. Nat Rev Dis Primers 2: 16028.
3. Atkinson FR (1937) Hereditary craniofacial dysostosis or Crouzon's syndrome disease. Med Press Circular 195: 118-124.
4. Golabi M (1984) Radiographic abnormalities of Crouzan syndrome. A survey of 23 cases. Proc Greenwood Genet Ctr 3: 102.
5. Cohen MM Jr (1986) Craniosynostosis: diagnosis, evaluation, and management. New York: Raven Press
6. Jeftha A, Stephen L, Morkel IA, Beighton P (2004) Crouzonoder-moskeletal syndrome. J Clin Pediatr Dent 28(2): 173-176.

ISSN: 2574-1241

DOI: 10.26717/BJSTR.2022.45.007181

Sandeep Aggarwal. Biomed J Sci & Tech Res