Narrow and duplicated internal auditory canal (IAC) is a rare congenital malformation of the temporal bone and usually combined with ipsilateral congenital sensorineural deafness. Congenital narrowing of IAC establishes a relative contraindication to cochlear implantation because it is associated with aplasia or hypoplasia of the vestibulocochlear nerve or cochlear branch. We report a 10-year-old boy with right sensorineural hearing loss. High resolution computed tomography (HRCT) reveals a narrow and duplicated IAC; the IAC is divided into an anterosuperior portion and posterior inferior portion by a bony plate. We present the CT finding and review the literatures in this statement.

CASE REPORT

A 10-year-old boy was told to have right side hearing impairment after hearing screening test of routine school medical exam. He denied any subjective hearing loss, otalgia, otorhoea or history of head injury. No abnormal speech development. The gross physical and otorhinolaryngological examination are normal. No any clinical complaint or sign of facial nerve abnormality.

Audiography revealed profound right sensorineural hearing loss and normal hearing on the left. The pure tone average is 7dB for left ear, but over 110 dB for right ear.

Coronal HRCT of the temporal bone showed a narrow and duplicated right IAC. The IAC is divided into an anterosuperior portion for facial nerve and a posteroimperior portion for vestibulocochlear nerve by a bony plate (Fig. 1). Three-dimensional reformatted CT medial to lateral view showed right IAC with double orifices (Fig. 2). Although unilateral SNHL was found in our young patient, he was not considered for surgical treatment and refused further MRI evaluation of temporal bone.

DISCUSSION

Narrow and duplicated IAC is a rare anomaly of the temporal bone, which commonly associated with
ipsilateral congenital sensorineural hearing loss. The first case of narrow and duplicated IAC was reported by Casselmen et al. in 1997 [5]. To date, only six cases of narrow IAC with duplication have been reported.

Development of the inner ear begins at 22 days of gestation. During the 8th week of gestation, the 7th and 8th cranial nerves become involved in the mesoderm, which forms the bony canal by chondrification and ossification [4]. Congenital narrowing of the IAC may lead to ipsilateral 8th cranial nerve hypoplasia or aplasia, but this hypothesis is controvertible due to the usually normal existence, development and function of the same-side facial nerve. When the 8th cranial nerve is aplasia or hypoplasia, the IAC fails to develop [8].

The vertical diameter of the IAC varies between 4 and 8 millimeters; if less than 2 mm, the IAC is considered stenotic [9, 10]. HRCT has a high sensitivity in examination of the bony structure of the IAC, but is also limited in assessing the neural elements in IAC. MRI is considered a good imaging modality to confirm the presence or non-existence of neural structure in narrowing IAC [5].

Assessment of the normal cochlear nerve is necessary when making a decision for implantation surgery [7]. Patients who do not have cochlea nerves cannot respond to the electric stimulation of cochlear implantation [11]. High-resolution submillimetric gradient-echo MRI is useful for further evaluation of the vestibulocochlear and facial nerve [12].

**Figure 1.** Coronal HRCT of temporal bone shows a narrow and duplicated right internal auditory canal (arrows). The internal auditory canalis divided into an anterosuperior portion and a posterosuperior portion by a bony plate structure.

**Figure 2.** Three-dimensional reformatted CT shows the duplicated right internal auditory canal with double orifices and the normal left internal auditory canal.
CONCLUSION

HRCT is a good imaging modality for temporal bone evaluation when the patient has congenital hearing impairment. If HRCT imaging reveals a narrow and duplicated IAC, it may indicate a hypoplastic or aplastic vestibulocochlear nerve, and the facial nerve can run through the upper separated canal. If a patient with congenital hearing impairment showing narrow IAC on CT image, and is considering acceptance of surgical cochlear implantation treatment, temporal bone MRI for vestibulocochlear nerve assessment is strongly recommended.

REFERENCES

狹窄雙層內耳道先天性畸形—
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狹窄雙層內耳道（IAC）是一種罕見的顳骨先天性畸形，它通常伴隨同側的先天性耳聾。人工耳蝸植入對先天性 IAC 狹窄來說是一項禁忌，因為它多合併有前庭耳蝸神經或耳蝸神經的發育不全。我們報告一位右耳聽覺喪失的 10 歲男童，其右側內耳道狹窄並分為上下兩部份管腔。顳骨高解析電腦斷層攝影的 3D 影像很清楚的顯示出先天性 IAC 的缺陷。在這份病例報告中，我們除了提出此例的 CT 影像外，也做文獻報告之回顧。