兩側先天性無耳蝸症之超短型MED-EL C40S多頻道人工耳蝸植入－病例報告

MED-EL C40S Conchlear Implantation in a Patient with Bilateral Congenital Cochlear Aplasia - Case Report

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摘要

先天性極重度聽力障礙合併耳蝸構造異常的病例在臨床上並不多見。文獻上，成功地對此類耳蝸畸形的病人執行多頻道人工蝸植入手術的報告亦寥寥可數。某些經篩選過的耳蝸結構異常病例，其多頻道人工耳蝸植入手術雖已逐漸被學者們認爲是切實可行的，但無耳蝸症的病例報告迄今仍付之闕如，而被視爲類手術的禁忌症。本院於1998年7月間，歷經此一13歲女童患有先天性極重度聽障合併兩側無耳蝸的病例。術前以高解像力顳骨電腦斷層攝影（high resolution CT of temporal bone）評估，發現兩側僅具聽小骨，前庭半規管而無耳蝸構造的發育。我們於術中移除蹬骨並在蹬骨足底板下之鼓室岬上之胚胎發育遺跡磨出一袋狀結構，並將MED-EL C40S超短型多頻道人工耳蝸植入。歷經術後30個月的持續追蹤，平均聽力一直穩定地維持在約35 dB HL的水準。這個特殊案例暗示著畸形耳蝸中可能仍有足夠數量的神經節細胞能讓人工耳蝸發揮作用。無耳蝸症及其它耳蝸畸形的人工耳蝸植入手術雖然有許多學理上的限制，但經審慎評估過的病例其成效仍是值得期待的。

Abstract

Various case reports describe patients with congenial profound deafness associate with cochlear malformations, but only a few report successful cochlear implantations in such patients. Multichannel cochlear implantation in selected patients with congenital malformed cochlear have gradually been shown to be feasible. To date, no reports describing successful cochlear implantation for cochlear aplasia have been published. Accordingly, cochlear aplasia remains an absolute contraindication to implantation. In July 1998, a successful multichannel cochlear implantation was performed in a 13-year-old girl with bilateral cochlear aplasia. Bilateral cochlear aplasia with normal ossicles and semicircular canals was shown on high resolution CT imaging of the temporal bone. The

incus was removed and the promontory below the foot plate where was the embryogenic remnants of the malformed cochlea was ground, and then the MED-EL C40S Cochlear Implant was implanted. Her average hearing remained around 35 dB HL during 30-mnoths of postoperative follow up. This special case suggests that there may be still sufficient ganglion cells within a malformed cochlea, that can allow cochlear implant to work. Despite theoretical contraindications to cochlear implantation for cochlear aplasia and other cochlear malformations, careful case selection could produce good results.