Cochlear implantation in case with KID syndrome

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Keratitis ichthyosis deafness (KID) syndrome is a rare, genetic disorder with eye problems, skin abnormalities and hearing loss. Less than 100 cases have been reported. It affects females slightly more often than males. Most of the reported cases were sporadic, but autosomal-dominant inheritance has been also reported. A few cases of KID syndrome caused by parental germline mosaicism for the GJB2 gene. The gene responsible for the KID-syndrome is called gap junction protein beta 2 (GJB2) and is located on the long arm of human chromosome 13 (13q11-q12). This gene encodes the structural protein “connexin-26”, which forms gap junction channels that connect neighboring cells and permit the exchange of small molecules and ions. The impairment of this connection and exchange may affect direct cell-to-cell communication in the skin and other tissues, such as the cornea and inner ear. In most cases hearing aids can’t be effectively used due to keratosis obturans of the external ear canal. Moreover, severe or profound hearing loss also limits the benefits gained from the conventional hearing aids. Still there are few cases reported in the literature about cochlear implantation is such patients.

Materials and methods
In 2017 cochlear implantation by using Cochlear Freedom CI24RE 512 (522) implant was performed in 3,5 years old child from Turkmenistan. Our thought was that connexin-related hearing loss is a good disorder for rehabilitation by cochlear implants.

Preoperatively
In order to minimize the clinical manifestations of the disease on the part of the skin, dermatological therapy has been held and included starch baths, keratolytic ointments, high doses of water-soluble and fat-soluble vitamins, especially retinol and tocopherol.

Intraoperatively
The surgery had no additional features. The skin was thick enough and an intradermal suture was used to close the wound. The operation has been held under general endotracheal anesthesia.

Postoperative care
The patient stayed in the hospital for 3 days followed by outpatient monitoring. Special dressing was weared and parenteral antibiotic was given for 5 days. During fitting and wearing of the speech processor the weakest magnet was used.

Results and conclusion
No complications were obtained during and after surgery. Implant fitting has been held in one month after implantation. Hearing tests had the good results. Cochlear implantation in KID syndrome can be save and effective.