

Abstract

Ehlers–Danlos syndrome (EDS) is a heterogeneous group of multisystem heritable disorders of connective tissue. Clinical features include excessive skin elasticity, excessive joint mobility, and increased tissue fragility. Few studies have been published on the otologic and audiological outcomes of patients with EDS.

The aim of the study is to present the results of stapedotomy in a long-term follow-up in a patient with EDS and to review the literature on the diagnostic process and otolaryngological management in this group of patients.

Rapport de Cas

- Currently, a 23-year-old woman with EDS, at the age 12, was admitted to the Department of Oto-Rhino-Laryngosurgery of the Institute of Physiology and Pathology of Hearing due to bilateral hearing loss
- Since early childhood, there were chronic upper respiratory tract infections and periodic inflammation of the ears with effusion. Physical examination with video otoscopy revealed a bilateral ventilation tube. Pure-tone audiometry confirmed bilateral mixed hearing loss: moderate in the right ear and severe in the left ear.

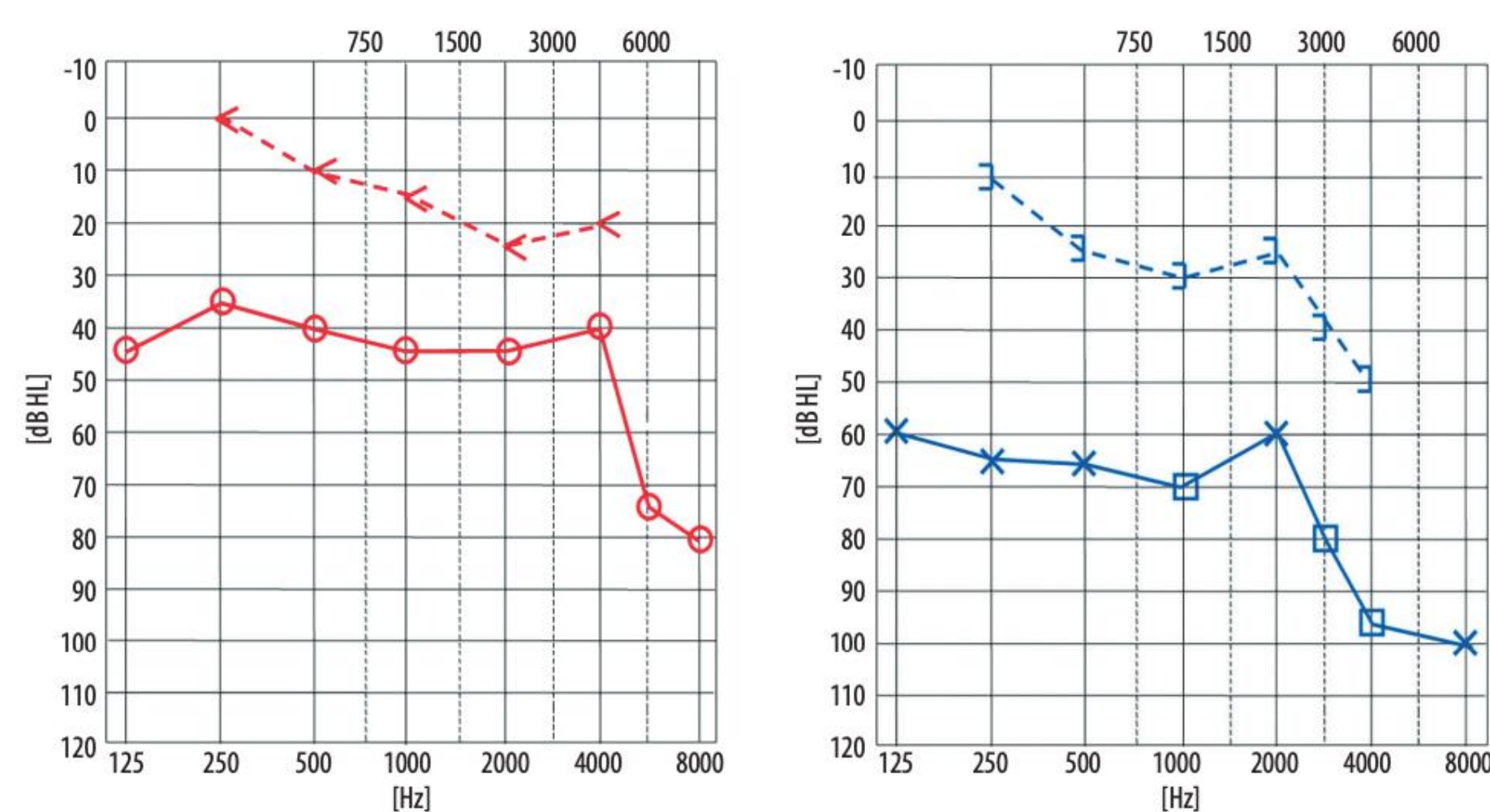


Figure 1. The patient's pure tone audiometry test results. Red: right ear; blue: left ear.

- The patient underwent bilateral stapedotomy. Intraoperatively, altered anatomical conditions within the middle ear indicating a malformation were found.

Résultats

During treatment at the Institute, a congenital malformation of the ossicles was identified in the patient. **Early post-operative pure tone audiometry confirmed significant hearing improvement following bilateral stapedotomy.** Importantly, no excessive intraoperative bleeding or abnormalities in the post-operative wound healing process were observed. **However, long-term follow-up revealed further progression of hearing loss, including sensorineural components.**

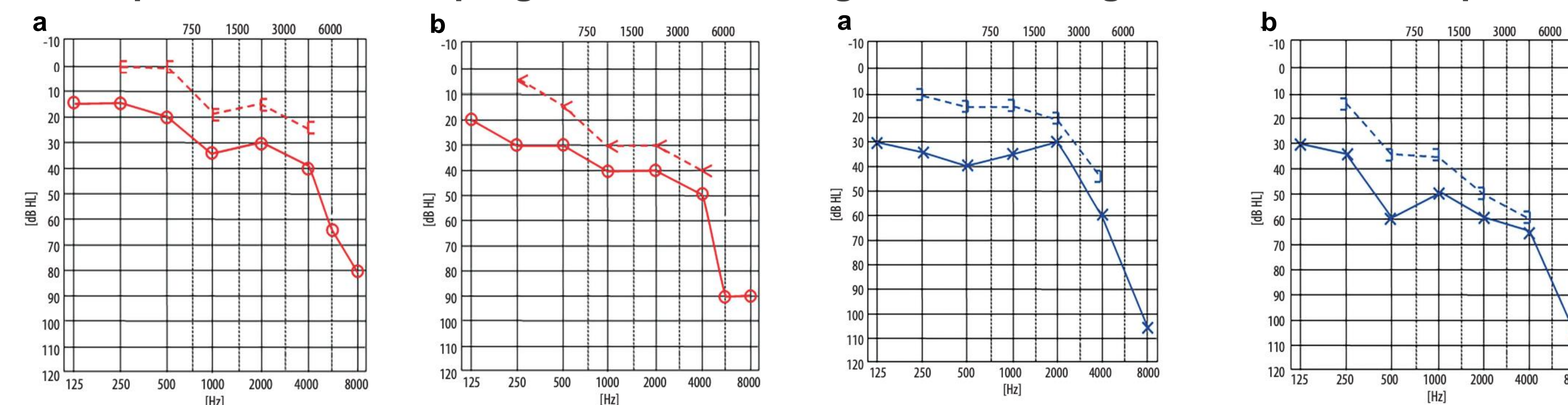


Figure 2. The patient's pure tone audiometry test results; a) 7 months after surgery, b) 2 years after surgery Red: right ear; blue: left ear.

Conclusion

There are single reports of EDS patients diagnosed with hearing loss. Because of the risk of intra- and postoperative complications based on heritable disorders of connective tissue, the authors postulate conservative treatment and **any surgery should be performed with extreme caution.**

Our patient's results showed that stapedotomy can provide hearing improvement, but surgery is more difficult due to significantly altered anatomical conditions.

Références

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