

# UNILATERAL HEARING LOSS AS A MAIN CLINICAL PRESENTATION OF CHIARI I MALFORMATION

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## INTRODUCTION

Chiari I malformation is a radiological diagnosis with a wide and varying clinical presentation, but most frequently presents with occipitocervical headache. Sensorineural hearing loss is rarely the main symptom. Possible theories of hearing loss in Chiari malformations are compression or traction of the vestibulocochlear nerve or the cochlear nuclei. Other theories include ischemia of the cochlear nuclei by the posterior inferior cerebellar artery (PICA) or its branches. Traction of the nerve does not seem explanatory since there's no brainstem herniation in Chiari I. The reversibility of the hearing loss makes ischemia less likely.

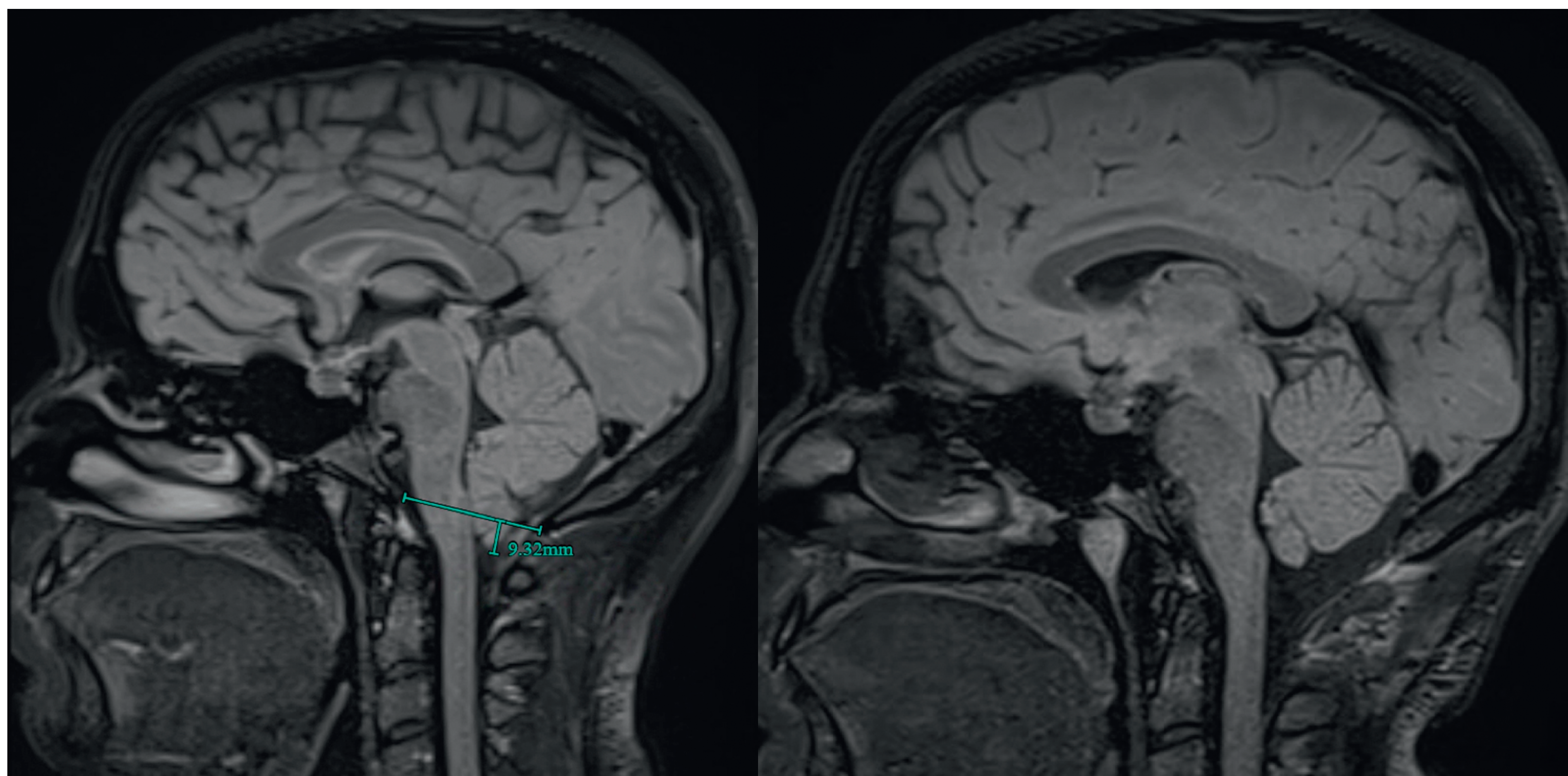


Figure 1: Preoperative MRI (T2 FLAIR) showing tonsillar herniation >9mm with compression of medulla oblongata. There was no hydrocephalus or syringomyelia.

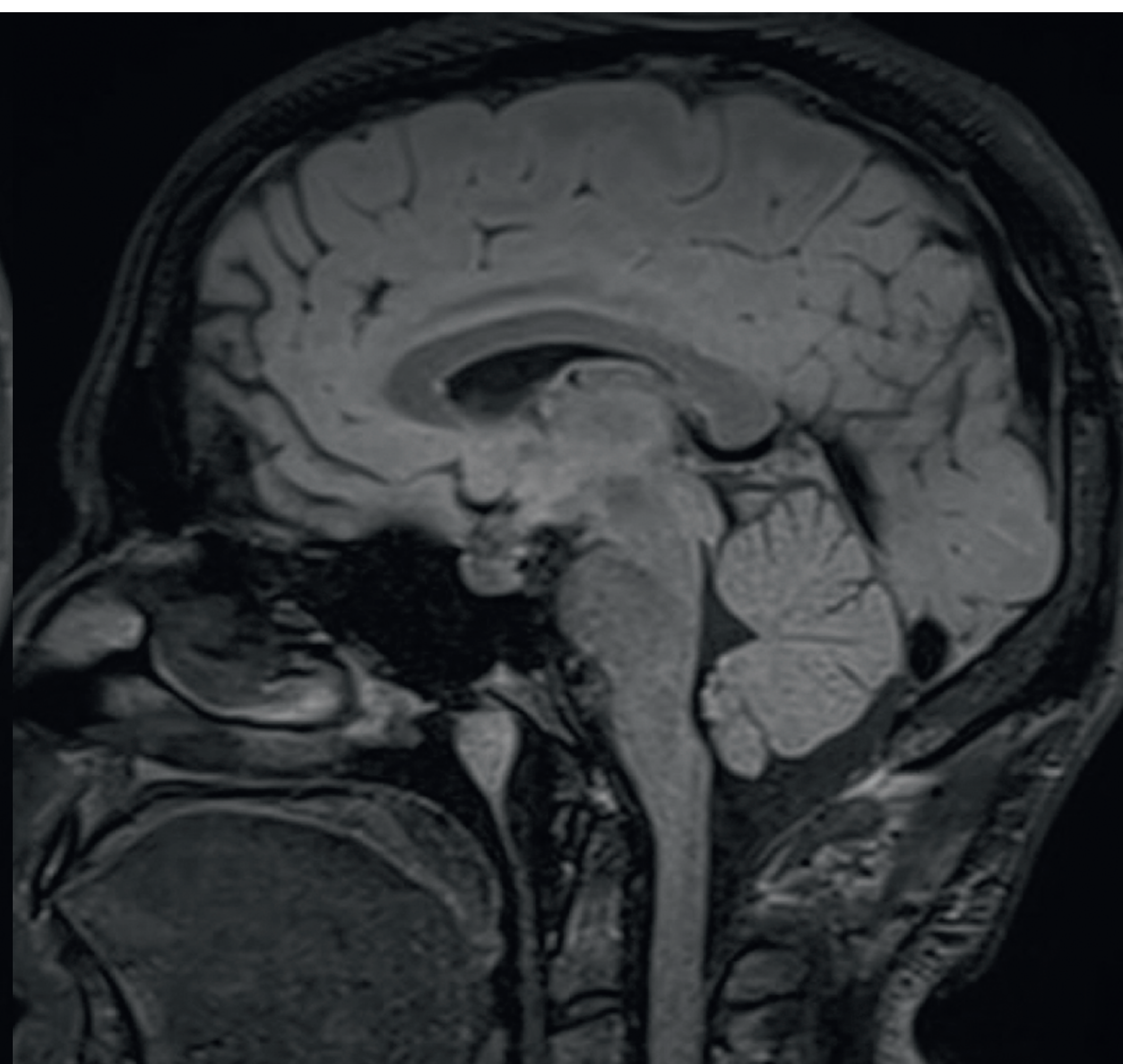


Figure 2: Postoperative MRI (T2 FLAIR) showing adequate decompression without sign of cerebrospinal fluid leakage.

## MATERIAL AND METHODS

We describe a 28-year-old patient presenting with rapid progressive unilateral hearing loss because of Chiari I malformation (Figure 1). Patient underwent suboccipital craniectomy with C1 laminectomy followed by tonsillar coagulation and fascia lata duraplasty.

## RESULTS

MRI scan showed adequate decompression without cerebrospinal fluid leakage (Figure 2). The unilateral hearing loss completely disappeared. This was confirmed with a pre- and postoperative audiogram (Figure 3).

## CONCLUSION

Chiari I malformation may present with sensorineural hearing loss as a main symptom. The hearing loss responded very well to fossa posterior decompression.

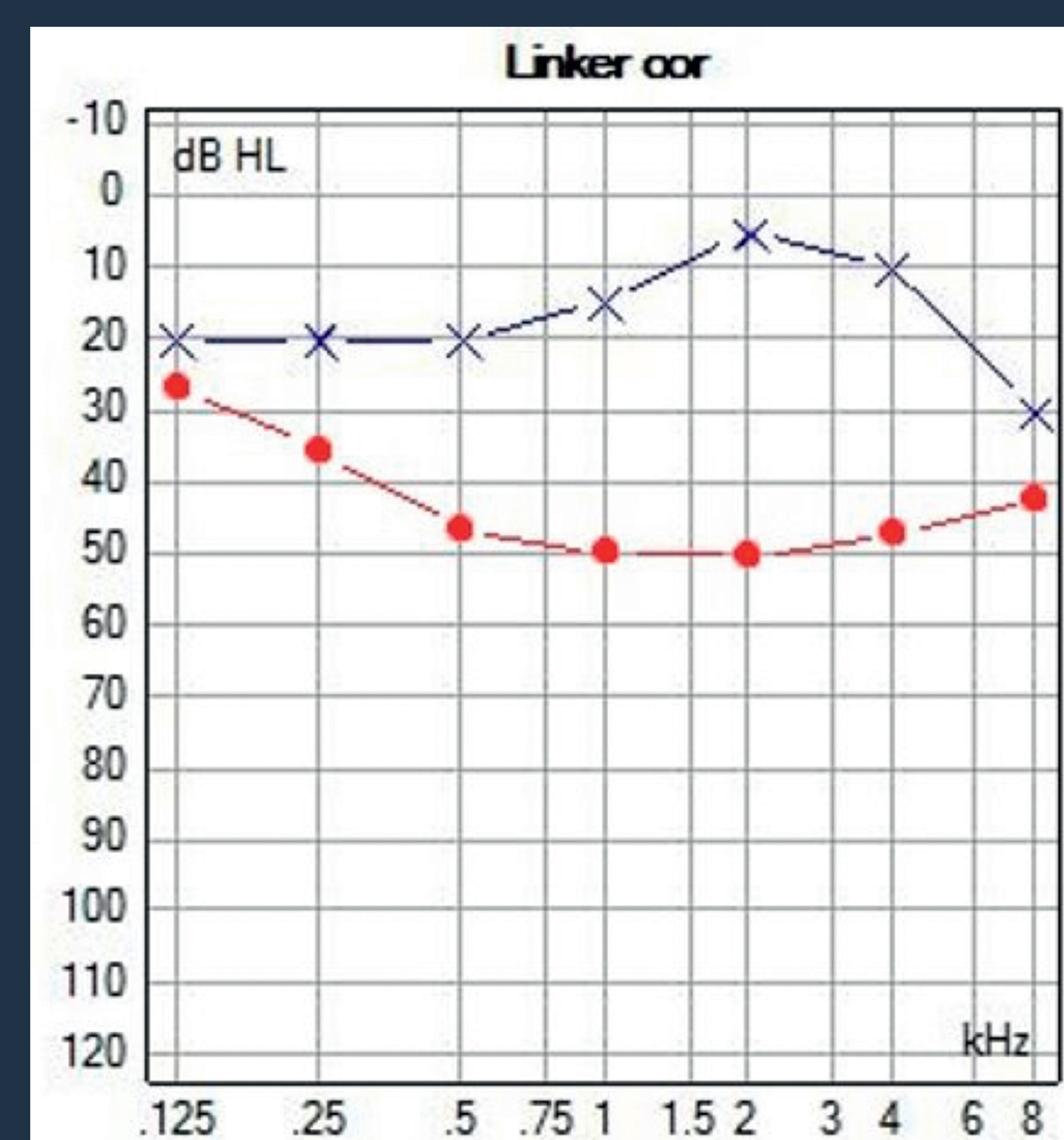


Figure 3: Pre- (dot) and postoperative (cross) audiogram showing normalization of the hearing loss, except for a subclinical remnant for the highest frequencies.