# Ossificação do tendão do estribo: Significado do ossículo de Paaw

# Stapedius tendon ossification: Meaning of the ossicle of Paaw.

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*ABSTRACT*

*Introduction: Stapes tendon ossification is an uncommon ossicular congenital malformation with autosomal dominant inheritance. Globular supernumerary sesamoid bone within the stapes tendon was described by several anatomists. We have found a fixed stapes by a bony bridge between pyramidal eminence and stapes head keeping the habitually morphology and consistency of the stapedial tendon.*

*Material and method: 40 year old female patient presented with progressive bilateral hearing loss history with similar family history. We present clinical examination, complementary studies, treatment and course.*

*Discussion: Patients with a conductive or mixed hearing loss, normal or low compliance, and acoustic reflex with inverted pattern in 2000 Hz, CT images can be diagnostic in 25-67 % of the cases, facilitating treatment planning. A linear image attenuation of soft tissues in CT from the pyramidal eminence to the stapes is suggestive of a congenital ossification of the stapedial tendon. A small globular or rounded image inside the stapes tendon corresponds to the existence of Paaw bone. Elimination of the bony bridge preserving the integrity of the ossicular chain and its tendons restores the problem of hearing loss.*

*Keywords: Stapedius tendon; Hearing loss, conductive; ossicular chain*

## INTRODUCTION

Familial conductive hearing loss is a rare condition that can be restored with an adequate diagnosis. Ossification of the stapes tendon is an uncommon ossicular congenital malformation by conversion of the interhyaline tissue (future stapedial tendon) into precartilaginous tissue leading to the stapes fixation[1,2]. The existence of a globular supernumerary sesamoid bone within the stapes tendon was described by several anatomists. Although now recognized to represent the lenticular process being part of the incus, its existence as a separate bone was a subject of controversy until 20th century[3]. We describe a case with fixed stapes by a bony bridge between pyramidal eminence and stapes head keeping the habitually morphology and consistency of the stapedial tendon.

## CLINICAL CASE

40-year-old female patient presented with progressive bilateral hearing loss history with similar family history. Otoscopy showed normal external ear and tympanic membrane. Bilateral Rinne test was negative and no lateralization was found at Weber test. Tympanometry showed a low compliance and bilateral absence of acoustic reflex, except at 500 and 2000 Hz in the right ear with inverted pattern; Audiometry revealed bilateral moderate mixed hearing loss with involvement of low and conversational frequencies. Average air conduction threshold in the right ear was 57 dB and 55 dB in the left ear, and bone conduction threshold average was 33 and 31 dB respectively. Maximum intelligibility threshold in speech audiometry was 70 dB in the right ear and 60 dB in the left ear.

Axial CT sections showed a linear bone density area extending from pyramidal eminence to stapes head in a right ear. (Figure 1).

We performed an exploratory tympanotomy of the right middle ear. Tympanomeatal flap was lifted up. Partial atticotomy was performed and we observed stapedial fixation by bony bridge between stapes head and pyramidal process. We found a linear prolongation with globular morphology of the tendon between pyramidal eminence and stapes head. The long process of the

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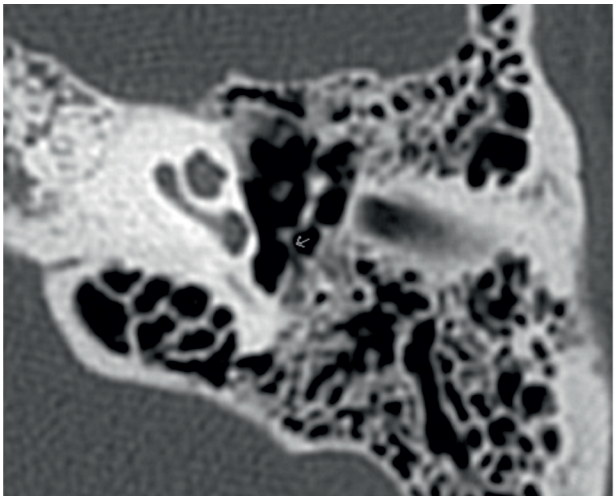
**FIGURE 1**
Coronal CT Scan. Arrow points a soft tissue linear image from piramidal eminence to stapes head corresponding to stapes tendon ossification.

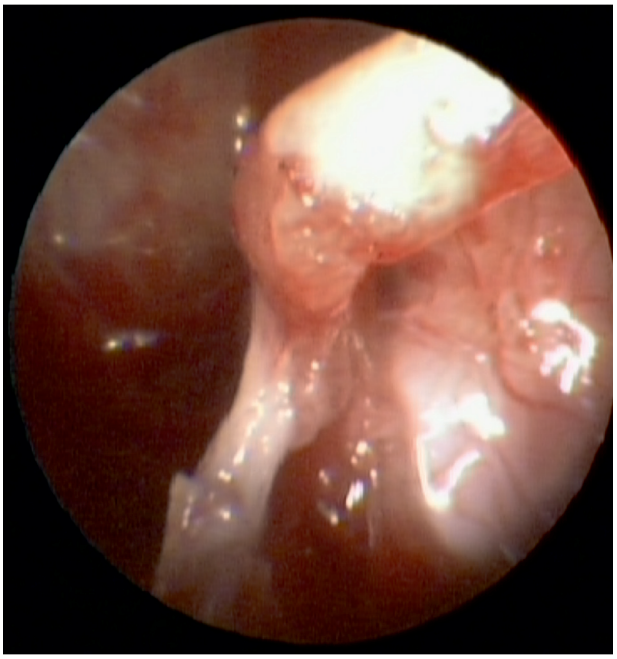


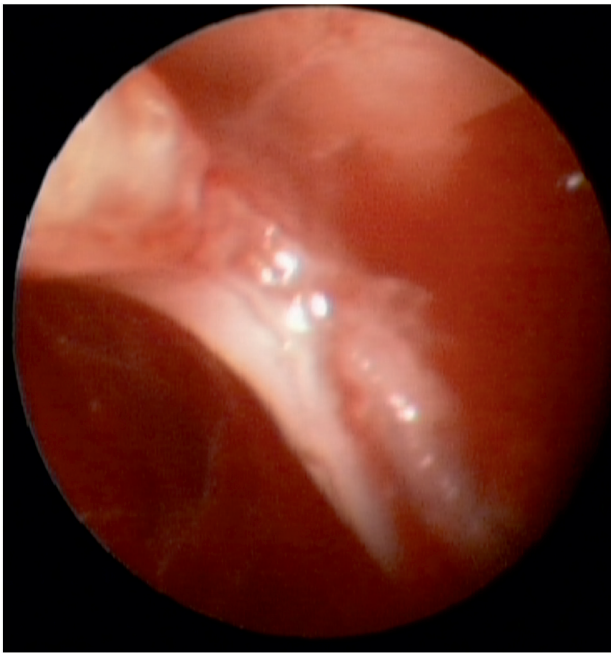
incus was discrete eroded in its caudal end, preserving the integrity of the lenticular process. (Figure 2). No otosclerosis focus was found in the footplate. Tympanic facial nerve was dehiscent covering partially the oval window. Drilling out the bony tract and keeping the tendon integrity it allows to reestablishing the normal mobility of the ossicular chain. Tragal perichondrium graft was used in the atticotomy area. Evolution was favorable with audiometry threshold improvement of 40 dB at 0.25 and 0.5 kHz, 30 dB at 1 kHz and 2 kHz and 20 dB at 4 kHz.

## DISCUSSION

Since the publication of the first cases of stapes tendon ossification by Schuknetcht and Trupiano in 1958, there have been several authors who have described the same malformation[1,2]. This is a congenital malformation with autosomal dominant inheritance pattern, more common in women, often with bilateral presentation. It is produced by conversion of interhialine tissue into precartilaginous inducing stapes fixation[1,2]. Stapes tendon exclusive impairment is due to its origin in the stem cells (mesenchymal or interhialine) of the second branchial arch which are different from those of the stapedius muscle and pyramidal eminence. The three structures stapedius muscle, stapes tendon and pyramidal eminence develops in the second arch from different histological cells blastemal cells, interhialine, and cartilaginous, respectively. Facial nerve dehiscence occurs by affecting the same mesenchymal stem cells which explains these association[1,2]. Diagnostic is supported by a conductive or mixed hearing loss with normal or low compliance, acoustic reflex typically inverted in 2000 Hz and bone conduction deterioration in the liquid test at 1KHz[2,4]. CT images allows us to confirm

**FIGURE 2**
Endoscopic image shows slightly osteitic long process of the incus and ossified stapes tendon with a globulous aspect in the adyacent portion to stapes head.





the diagnosis and surgical treatment planning[2,5,6]. CT 1 mm images can detect between 25-67 % of the cases. 65 % have a complete stapedius tendon ossification[6]. A linear area of soft tissue attenuation extending from pyramidal eminence to the posterior crura suggests congenital tendon stapedius ossification[6,7]. The ligament thickness is measured in axial images using as reference the anterior stapes crura and in coronal images the tensor tympani tendon, which are abnormal when they double in size. Tendon ossification

is confirmed by measuring the density, by comparing it with the density of the otic capsule. Density values of 400 HU or more were accepted as evidence of tendon ossification[6]. Conductive hearing loss with ossification ligaments detected by CT lead us to suspect this entity[6]. Differential diagnosis includes tympanosclerosis focus ruled out by absence of pathology in the middle ear; and otosclerosis, due to the presence of Carhart notch in both entities, discarded by the normal movility of the footplate[2,8]. In 1600 anatomists described the existence of Paaw bone, or "osseus globulus", " lenticular bone " or " fourth ossicle, " described as a small sesamoid rounded ossicle located inside the stapedial tendon which function was to reduce the stapedius tendon friction between adjacent mucous membranes of the middle ear. Its existence was discussed until the twentieth century, when it was accepted that it exists in some mammals, whereas in humans became more controversial and for several authors it represented the lenticular process[3]. Some of the descriptions made were partial ossification, which may mimic, as in this case, a fourth ossicle. The treatment is surgical, usually during exploratory tympanotomy. Removing the malformation by drilling the bone bridge, and at the same time preserving the integrity of the ossicular chain and tendons, provides good ossicular mobility and restores hearing within a normal range[2,3].

**Protecção de pessoas e animais**

Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

**Confidencialidade dos dados**

Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

**Conflito de interesses**

Os autores declaram não ter nenhum confito de interesses relativamente ao presente artigo.

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