The Journal of International Advanced Otology

J Int Adv Otol 2025; 21: 1–6 • DOI: 10.5152/iao.2025.241755

Case Report

A New Internal Acoustic Canal Anomaly, Fused Single Internal Acoustic Canal: A Case Report

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Cite this article as: Akşamoğlu M, Muluk NB, Şahan MH, Leblebici A, Tunç O. A new internal acoustic canal anomaly, fused single internal acoustic canal: A case report. *J Int Adv Otol.* 2025, 21(3), 1755, doi: 10.5152/iao.2025.241755.

The internal auditory canal (IAC) connects the inner ear to the posterior cranial fossa.

In this case report, the authors presented an 8-month-old patient with an IAC anomaly and bilateral deafness. The patient has hypotonia and epilepsy, and there is multifocal epileptiform activity on Electroencephalography (EEG). Severe (total) sensorineural hearing loss was detected bilaterally. Temporal bone computed tomography showed that the internal acoustic canal is localized as a single entity at the midline and bifurcates into right and left branches approximately ~4 mm beyond. Following branching, the diameter of each IAC measures approximately ~2 mm. On both sides, inner ear structures are located more medially than normal, positioned close to the midline and each other. Both cochleae undergo approximately 1.5 turns, classified as cochlear hypoplasia type III according to Sennaroğlu and Bajin's classification. Magnetic resonance imaging showed that in the anterior aspect of the pons, extending from the midline to the internal acoustic canal, a paired appearance, resembling a couple, of the VIII cranial nerve is observed in close proximity to each other. The pair of the VIII nerve seems to diverge at the entrance of the internal acoustic canal, and immediately thereafter, they bifurcate into branches believed to belong to the superior and inferior vestibular nerves on both sides, but the cochlear nerve cannot be discerned.

To the best of current knowledge, this is the first case in the literature as fused IAC.

KEYWORDS: cochlear turns, deafness, fused IAC, internal acoustic canal, 1.5 turns

INTRODUCTION

The internal auditory canal (IAC), also known as the "internal acoustic meatus," is situated within the temporal bone, connecting the inner ear to the posterior cranial fossa. Structures found within it comprise the "vestibulocochlear nerve (CN VIII), facial nerve (CN VII), the labyrinthine artery, and the vestibular ganglion."[1]

In this case report, an 8-month-old patient with an IAC anomaly, fused single internal acoustic canal, and bilateral deafness was presented.

CASE REPORT

The 8-month-old girl who has hypotonia and epilepsy was sent to cranial MRI in Gaziantep University, Faculty of Medicine, Radiology Department. In the Pediatric Neurology Department of an external center, there was a suspicion of congenital muscular dystrophy, but there is no diagnosis yet. They found multifocal epileptiform activity on EEG. In the Otorhinolaryngology Department of Gaziantep University, severe (total) sensorineural hearing loss was detected bilaterally by auditory brainstem response.

There is no need to obtain ethics committee approval, because this is a case report. However, the mother and father of the patient gave written permission to publish the clinical data and radiological views of the patient for scientific purposes.

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Received: October 4, 2024 • **Revision Requested:** November 5, 2024 • **Last Revision Received:** January 18, 2025 • **Accepted:** February 4, 2025 • **Publication Date:** May 28, 2025
Available online at www.advancedotology.org

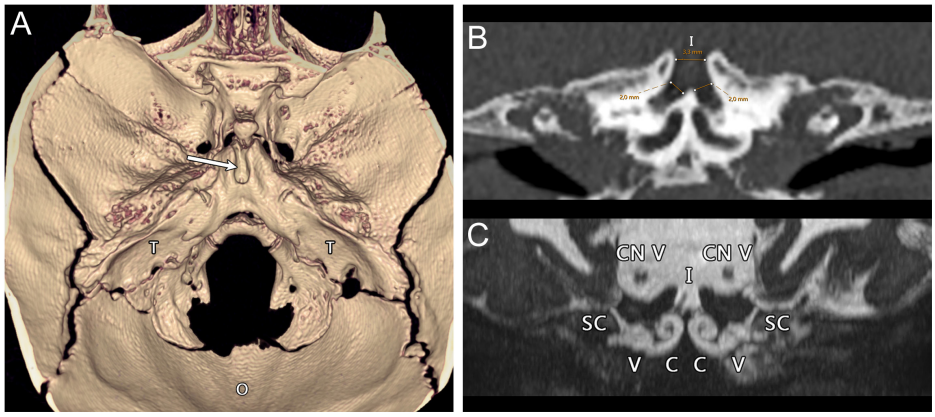

Figure 1. (A) Top view of the 3D surface rendering of temporal bone CT. (B) Coronal oblique view of temporal bone CT passing through the IAC level. (C) Coronal oblique MIP view passing through the IAC level of MRI. Abbreviations: Arrow, single internal acoustic canal in the midline; C, cochlea; CN V, cranial nerve V (trigeminal nerve); I, internal acoustic canal; O, occipital bone; SC, semicircular canal; T, temporal bone; V, vestibule.

Temporal Bone Computed Tomography Findings

The internal acoustic canal is localized as a single entity at the midline and bifurcates into right and left branches approximately ~4 mm beyond (Figure 1A-C). Following branching, the diameter of each IAC measures approximately ~2 mm (Figure 1B and C, 2A-C).

In both sides, inner ear structures are located more medially than normal, positioned close to the midline and to each other (Figure 2D-F).

Both cochleae undergo approximately 1.5 turns (Figures 2E and F, 3), classified as cochlear hypoplasia type III (Figures 2E and 3) according to Sennaroğlu and Bajin's classification.[2] On the right, basal, and middle turns are present, but the apical turn is not visualized (1.5 turns). On the left, the middle turn terminates after slight expansion. Although the appearance on the left resembles incomplete partition type II (IP-II), it was not classified as IP-II because the cochlear size is slightly smaller than normal. Although not always, a broad vestibular aqueduct (VA) is commonly associated with IP-II.[2] In the patient, the VA was of normal width (Figure 2E).

On the right, the cochlear aperture is stenotic, and the modiolus is hypoplastic. On the left, the cochlear aperture is atretic, and the modiolus is absent (Figure 4). This is considered cochlear nerve agenesis and/or hypoplasia.

Both vestibules are dilated (Figures 2D-F and 3). Cystic dysplasia is present in both lateral semicircular canals (Figures 2D-F and 3).

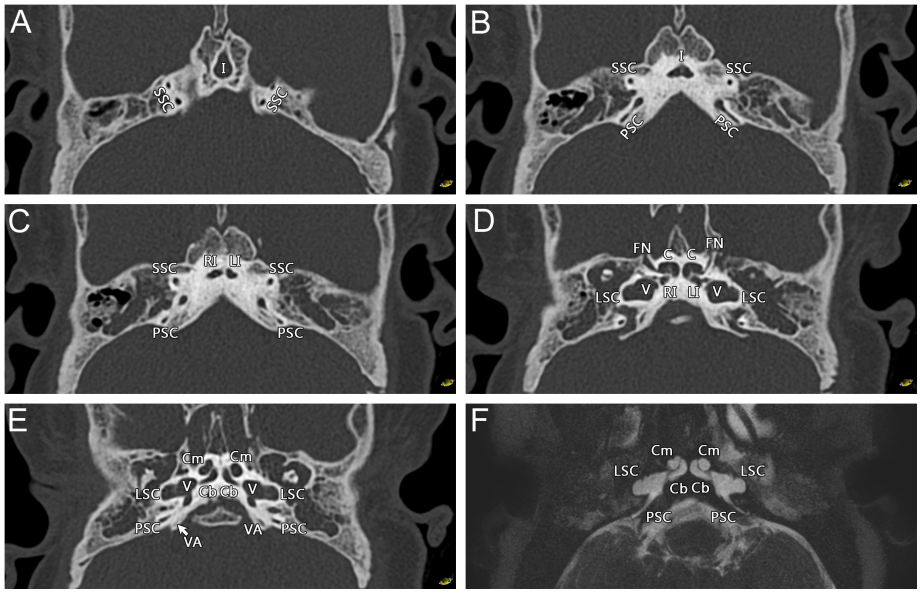

Figure 2. Serial axial images of temporal bone CT from superior to inferior: (A) Level where the internal acoustic canal (IAC) is observed singly in the midline. (B) Level where the IAC is divided into right and left. (C) Levels of the right and left IAC. (D) Vestibular level. Vestibules are dilated bilaterally, and lateral semicircular canals appear cystically dysplastic. The facial nerve canal is observed bilaterally but notably narrow on the left. (E) Cochlear level. Cochlear structures are nearly normal in size bilaterally, but the basal turns of the cochlea are slightly smaller. On the right, the middle turn is present but blind-ended, and the apical turn is not visualized (1.5 turns). On the left, the middle turn terminates after slight expansion. (F) Serial axial images of temporal bone CT from superior to inferior: MIP view of MRI passing through the cochlear level. Abbreviations: C, cochlea; Cb, cochlear basal turn; Cm, cochlear middle turn; CN V, cranial nerve V (trigeminal nerve); FN, facial nerve canal; I, internal acoustic canal; LI, left internal acoustic canal; LSC, lateral semicircular canal; PSC, posterior semicircular canal; RI, right internal acoustic canal; SSC, superior semicircular canal; V, vestibule; VA, vestibular aqueduct.

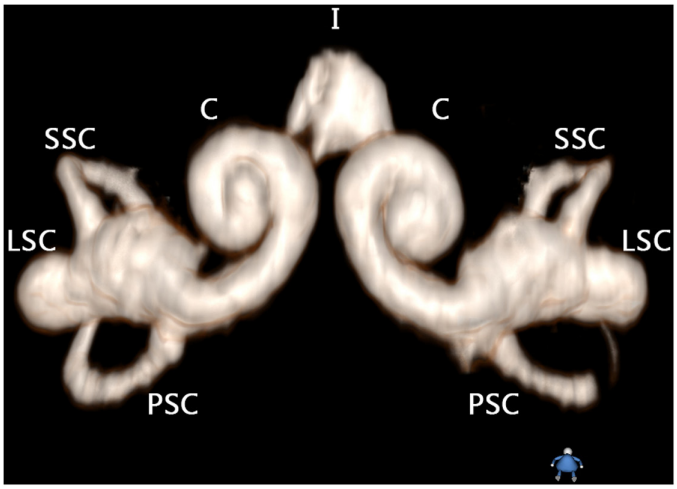

Figure 3. Coronal 3D view of the MR image. Abbreviations: C, cochlea; I, internal acoustic canal; LSC, lateral semicircular canal; PSC, posterior semicircular canal; SSC, superior semicircular canal.

Superior semicircular canals on both sides show bone dehiscence in the superior aspect.

There is a facial nerve canal on both sides, although it is narrower on the left side (Figure 2D).

Magnetic Resonance Imaging Findings

In the anterior aspect of the pons, extending from the midline to the internal acoustic canal, a paired appearance, resembling a couple, of the VIII cranial nerve is observed in close proximity to each other (Figure 5).

The pair of the VIII nerve seems to diverge at the entrance of the internal acoustic canal, and immediately thereafter, they bifurcate into branches believed to belong to the superior and inferior vestibular nerves on both sides, but the cochlear nerve cannot be discerned (Figure 6A and B).

At the level of exit from the pons of the VIII cranial nerve, on both sides of this nerve, 2 very thin-caliber (~0.4 mm) nerve appearances are observed, which appear blurred and hence could be interpreted as facial nerves. The facial nerve, presumably due to its thin caliber,

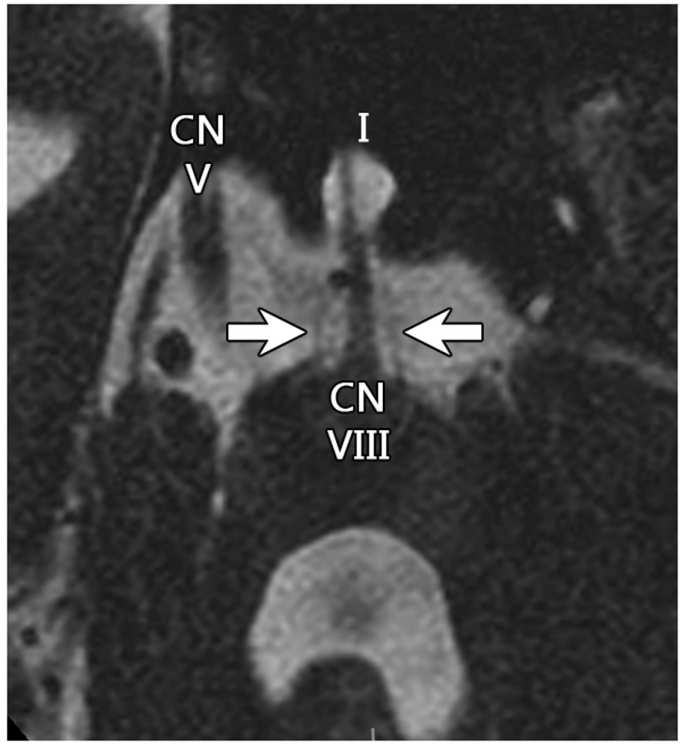

Figure 5. Axial oblique view passing through the exit level of cranial nerve VIII from the pons. Abbreviations: CN V, cranial nerve V (fifth cranial nerve); CN VIII, cranial nerve VIII (eight cranial nerve); I, internal acoustic canal.

cannot be distinguished even at the level of the internal acoustic canal (Figure 5).

Other Findings

Bilateral ossicles of the ears are observed to be normal.

In the sections covered by the examination:

The volume of the pons is decreased, appearing hypoplastic. The posterior fossa is shallow.

The fourth ventricle is narrowed inferiorly. The third ventricle and both lateral ventricles are dilated.

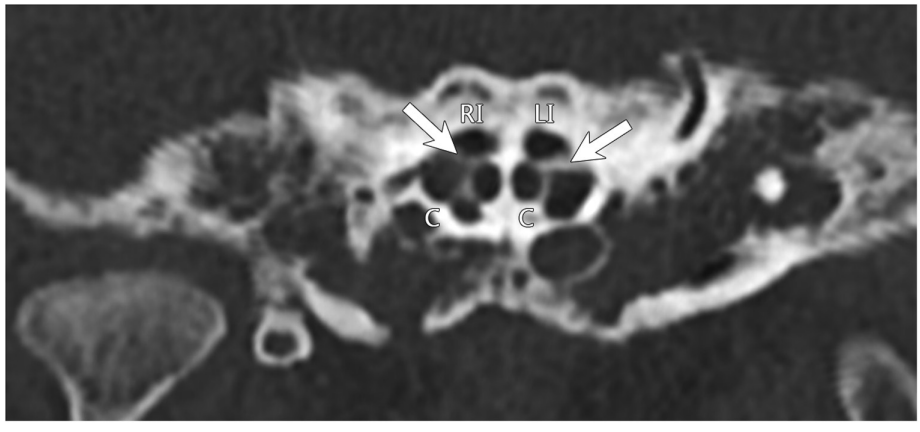

Figure 4. Coronal oblique view passing through the cochlear aperture level of temporal bone CT. Abbreviations: Arrows, cochlear aperture region; C, cochlea; LI, left internal acoustic canal; RI, right internal acoustic canal.

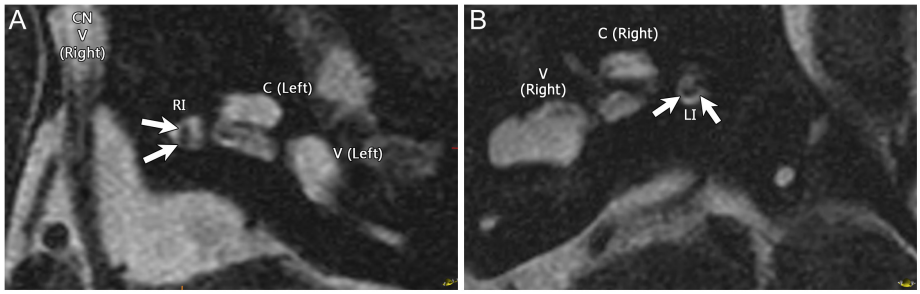

Figure 6. Perpendicular reformat images of (A) the right and (B) left internal acoustic canal (IAC) route after branching into right and left branches. Two nerve branches (superior and inferior vestibular nerves) emerge from CN VIII on both sides. Abbreviations: C, cochlea; CN V, cranial nerve V (fifth cranial nerve); LI, left internal acoustic canal; RI, right internal acoustic canal; V, vestibule.

The Brain and Cranial Anomalies Detected on Magnetic Resonance Imaging

- The pons appears hypoplastic with reduced volume.
- The posterior fossa is shallow.
- The fourth ventricle is narrowed inferiorly, while the third and both lateral ventricles are dilated.
- On the right, the intracranial segment of the internal carotid artery (IAC) appears hypoplastic.
- Both ICAs are located near the midline.
- The body of the sphenoid bone is hypoplastic.
- The retroorbital distance is shortened.
- The pituitary fossa is in its normal position, but instead of the commonly observed oval cavity appearance, it is seen as a wide-angled triangular shape with the base upwards (Figure 7).
- The clivus is in a vertical position, and basilar kyphosis is present (Figure 8).
- No anomalies were detected in the corpus callosum or other midline structures.

Follow-Up Information for the Patient

During the MRI scan, the patient was already dyspneic. Shortly after the scan, she was admitted to the intensive care unit due to aspiration pneumonia. Later, she was discharged. A cochlear implant was not performed because it was determined that the patient lacked the 8th cranial nerve, and a brainstem implant was planned instead. However, it has not been placed yet. Neurological treatment has not been planned for the patient in thehospital, and the patient has not attended the follow-up appointments.

DISCUSSION

"The vestibulocochlear nerve" usually lies posterior to "the facial nerve" within the IAC. Approximately 3-4 mm from the fundus in the lateral section of the canal, "the cochlear and vestibular nerves" merge into a single bundle. Close to the fundus, the vestibular part divides into superior and inferior branches.[3] The superior branch innervates 3 structures: "the superior and lateral semicircular canals, and the utricle." Conversely, the inferior branch serves the remaining vestibular components, including "the posterior semicircular canal and the saccule." Efferent fibers from both branches meet in the IAC at the vestibular ganglion, often referred to as the "Scarpa ganglion." Notably, this ganglion reaches its full size early in postnatal development, typically within the first week.[4]

In the case report, an IAC anomaly causing deafness was detected. Temporal bone CT showed that the internal acoustic canal is localized as a single entity at the midline and bifurcates into right and left branches approximately ~4 mm beyond. Following branching, the diameter of each IAC measures approximately ~2 mm. On both sides, inner ear structures are located more medially than normal, positioned close to the midline and to each other. Both cochleae undergo approximately 1.5 turns, classified as cochlear hypoplasia type III according to Sennaroğlu and Bajin's classification.[2] Magnetic resonance imaging showed that in the anterior aspect of the pons,

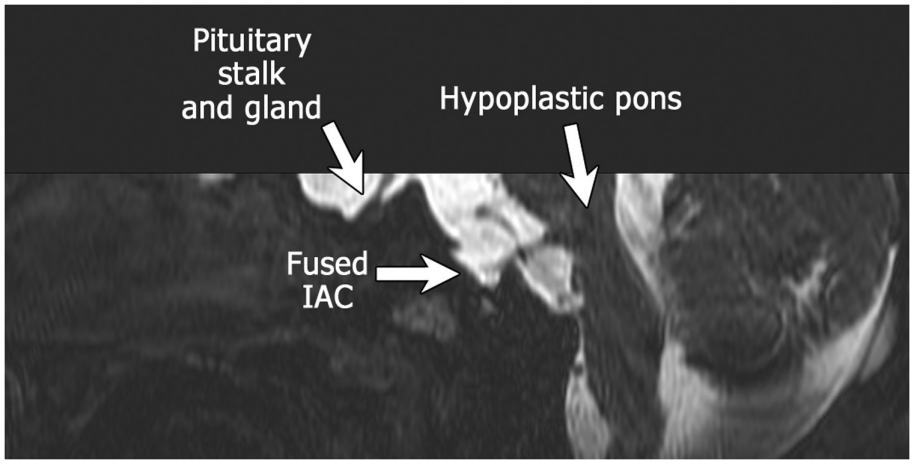

Figure 7. Midsagittal reformatted image of the T2W 3D-DRIVE sequence. Hypoplastic pons, fused internal acoustic canal (IAC), pituitary stalk, and gland are observed.

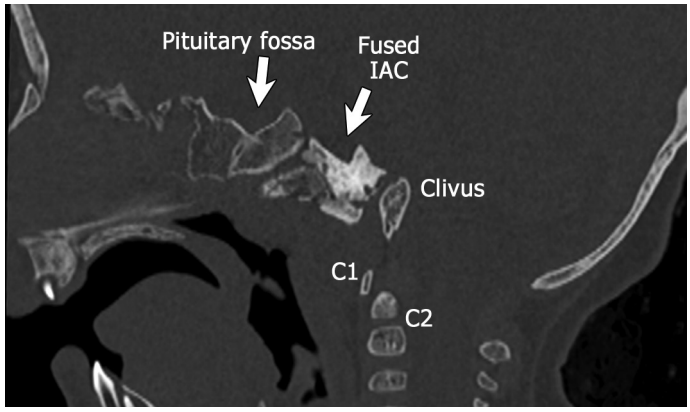

Figure 8. Midsagittal reformatted image of the temporal CT. Fused internal acoustic canal (IAC), atypical triangle-like pituitary fossa, upright clivus causing basilar kyphosis, and the relationship of the clivus with the upper cervical vertebrae are observed.

extending from the midline to the internal acoustic canal, a paired appearance, resembling a couple, of the VIII cranial nerve is observed close to each other. The pair of the VIII nerve seems to diverge at the entrance of the internal acoustic canal, and immediately thereafter, they bifurcate into branches believed to belong to the superior and inferior vestibular nerves on both sides, but the cochlear nerve cannot be discerned. As per current knowledge, this is the first case in the literature as fused IAC. However, a narrow, duplicated, or triplicated IAC is reported as a very rare anomaly.[5-7]

Since this case is presumed to be a type of craniofacial malformation, the position of this case within craniofacial malformations was discussed[8]:

Craniosynostosis[8]: Apert syndrome presents with "craniosynostosis, midface hypoplasia, and symmetric syndactyly of hands and feet." Features like "hypertelorism (widely spaced eyes), proptosis (bulging eyes), and downward-slanting palpebral fissures" are common in several craniosynostotic conditions.[9] Both Crouzon and Apert syndromes also involve a shortened bony orbit, reduced orbital and soft-tissue volume, and increased globe volume. In Crouzon syndrome, the middle cranial fossa is displaced anteriorly and inferiorly, shortening the orbit further.[10] A decreased retro-orbital distance, similar to these syndromes, was exhibited by the patient. In Crouzon syndrome, additional cranial deformities like brachycephaly, trigonocephaly, or oxycephaly may occur, often linked to sphenoid hypoplasia.[10,11] This sphenoid hypoplasia is also observed in Cleidocranial dysplasia, an autosomal dominant disorder caused by mutations in the RUNX2 gene.[12] Sphenoid hypoplasia was also presented by the patient.

Encephalocele (which may be accompanied by "absent corpus callosum, clefts, Dandy–Walker, Arnold–Chiari malformations, ectrodactyly, and hypothalamic-pituitary dysfunction"[8]) can lead to significant posterior fossa and hindbrain abnormalities, including cerebellar herniation, absence of the "cerebellum, and associated issues like hydrocephalus, syrinx, encephalocele, or spinal malformations."[13] In the patient, the pons appear hypoplastic with reduced volume, and the posterior fossa is shallow; no anomalies were detected in the corpus callosum or other midline structures. No encephalocele was detected in the patient.

Syndromes like Noonan syndrome, which is an inherited disorder with a variety of phenotypic expressions, often change over time. The most consistent features include wide-set eyes, low-set ears, short stature, and pulmonic stenosis.[14] In this case, anomalies in facial appearance were not detected.

The patient has similarities to craniosynostosis and a negative association with other craniofacial malformation-causing diseases such as encephalocele and Noonan syndrome.

In conclusion, the first case of fused IAC is presented in the literature. Sphenoid hypoplasia and decreased retro-orbital distance in this case may be related to craniosynostosis.

Availability of Data and Materials: All data for this study is presented in the paper.

Informed Consent: Written informed consent was obtained from the patients's parents who agreed to take part in the study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – M.A., N.B.M., M.H.Ş., A.L., O.T.; Design – M.A., N.B.M., M.H.Ş., A.L., O.T.; Supervision – M.A., M.H.Ş.; Resources – M.A., N.B.M., M.H.Ş., A.L., O.T.; Materials – M.A., M.H.Ş., A.L., O.T.; Data Collection and/or Processing – M.A. M.H.Ş., A.L., O.T.; Analysis and/or Interpretation – M.A., N.B.M., M.H.Ş., A.L., O.T.; Literature Search – M.A., N.B.M., M.H.Ş., A.L., O.T.; Writing – N.B.M., M.A.; Critical Review – M.A., N.B.M., M.H.Ş., A.L., O.T.

Declaration of Interests: The authors have no conflicts of interest to declare.

Funding: The authors declared that this study received no financial support.

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