

ORIGINAL ARTICLE

## Defect of the Bony Roof in the Superior Semicircular Canal and Its Clinical Implications<sup>☆</sup>

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### KEYWORDS

Bony defect;  
Superior semicircular  
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### Abstract

**Introduction and objective:** The aetiology of the superior semicircular canal dehiscence is currently unknown. Our objective was to analyse and discuss different hypotheses about the origin of this pathology.

**Methods:** In this study performed on 295 temporal bones, one case of partial alteration of the bony roof in the right superior semicircular canal was described from the anatomical and radiological points of view, and compared with the temporal bone on the other side.

**Results:** Macroscopically, the superior semicircular canal shows deterioration in the bony roof, which consists exclusively of the inner or endosteal layer that separates the canal from the superior semicircular conduct.

The Pöschl plane reconstruction showed a whole bony roof, but its thickness decreased from the canal curvature to the defect (from 0.6 to 0.3 mm).

**Conclusion:** The presence of partial defects in the bony roof of the superior semicircular canal with absence of the external and middle layers, besides its lesser thickness, makes the canal susceptible to suffering a second event. This could produce its fracture and a dehiscence.

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### PALABRAS CLAVE

Defecto óseo;  
Canal semicircular  
superior;  
Hueso temporal  
humano

### Defecto de cobertura del canal semicircular superior y su implicación clínica

#### Resumen

**Introducción y objetivo:** En la actualidad se desconoce la etiología del síndrome de dehiscencia del canal semicircular superior. Nuestro objetivo es analizar y discutir las distintas hipótesis sobre el origen de dicha patología.

**Métodos:** En un estudio sobre 295 temporales se describe desde el punto de vista anatómico y radiológico un caso de una alteración parcial de la cobertura ósea del canal semicircular superior derecho y se compara con el temporal del otro lado.

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**Resultados:** Macroscópicamente el canal semicircular superior exhibe un deterioro de su cubierta ósea, constituida exclusivamente por su capa más interna o endostal que separa el canal del conducto semicircular superior.

La reconstrucción en el plano Pöschl demuestra que aunque la cubierta está íntegra, su grosor disminuye a lo largo de la curvatura del canal hasta el defecto (0,6 a 0,3 mm).

**Conclusión:** La presencia de defectos parciales en la cobertura ósea del canal semicircular superior con ausencia de sus capas externa y media, sumado a su menor grosor, hacen susceptible al canal para sufrir un segundo evento que podría causar su rotura pudiéndose convertir en una dehiscencia.

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## Introduction

In 1998, Minor et al.<sup>1</sup> showed that some vestibular cases frequently associated with hearing loss were due to superior semicircular canal dehiscence. This syndrome was therefore referred to as superior semicircular canal dehiscence syndrome, in reference to the clinical case associated with the lack of bone coverage of this canal at the level of the middle cranial fossa.

Following this, there have been numerous publications describing its anatomical<sup>2,3</sup> and radiological<sup>4-8</sup> incidences, and even mixed studies examining both the anatomical and the radiological incidences, showing a remarkable lack of agreement between both.<sup>9</sup>

The aetiopathogenesis of this dehiscence is unknown. Various hypotheses have been postulated, including alteration of postnatal development of the middle and outer layers of the bone canal,<sup>2</sup> incorrect position of the primitive otocyst,<sup>3</sup> a bone reorganisation defect in the canal during the prenatal period,<sup>10</sup> genetic factors with mutation of the COCH gene,<sup>11</sup> thinning of the canal cover,<sup>12</sup> breakage of the canal by head trauma and increased intracranial pressure.<sup>13</sup>

## Material and Methods

This was a study of the presence of superior semicircular canal dehiscence in temporal bones (295) belonging to the ossuary of the Human Anatomy and Histology Department of the School of Medicine at the University of Zaragoza.

We studied the isolated temporal bones of a 6-year-old boy who suffered a defect in the cover of this canal, from both the anatomical and the radiological standpoints.

The radiological study was carried out with a multi-section spiral computerized tomography device (Philips Brilliance 6).

The imaging protocols for the acquisition and formatting of images were as follows: collimation 2×0.6 mm, section width 0.65 mm, section increase 0.32 mm, rotation time 0.75 s, pitch 0.38, 120 kV, 300 mAs, matrix 1024×1024, field of view 180 mm, reconstruction width 0.5 mm and reconstruction increase 0.5 mm.

Reconstructions were carried out in the coronal plane, as well as in the plane of the superior semicircular canal in each temporal bone. The raw data were reconstructed using a bone algorithm.

## Results

The descriptive study of both temporal bones allowed us to observe the partial alteration of the cover of the superior semicircular canal on the right side (Fig. 1) while the left (Fig. 1c) was completely normal. Both bones showed a large arched space.

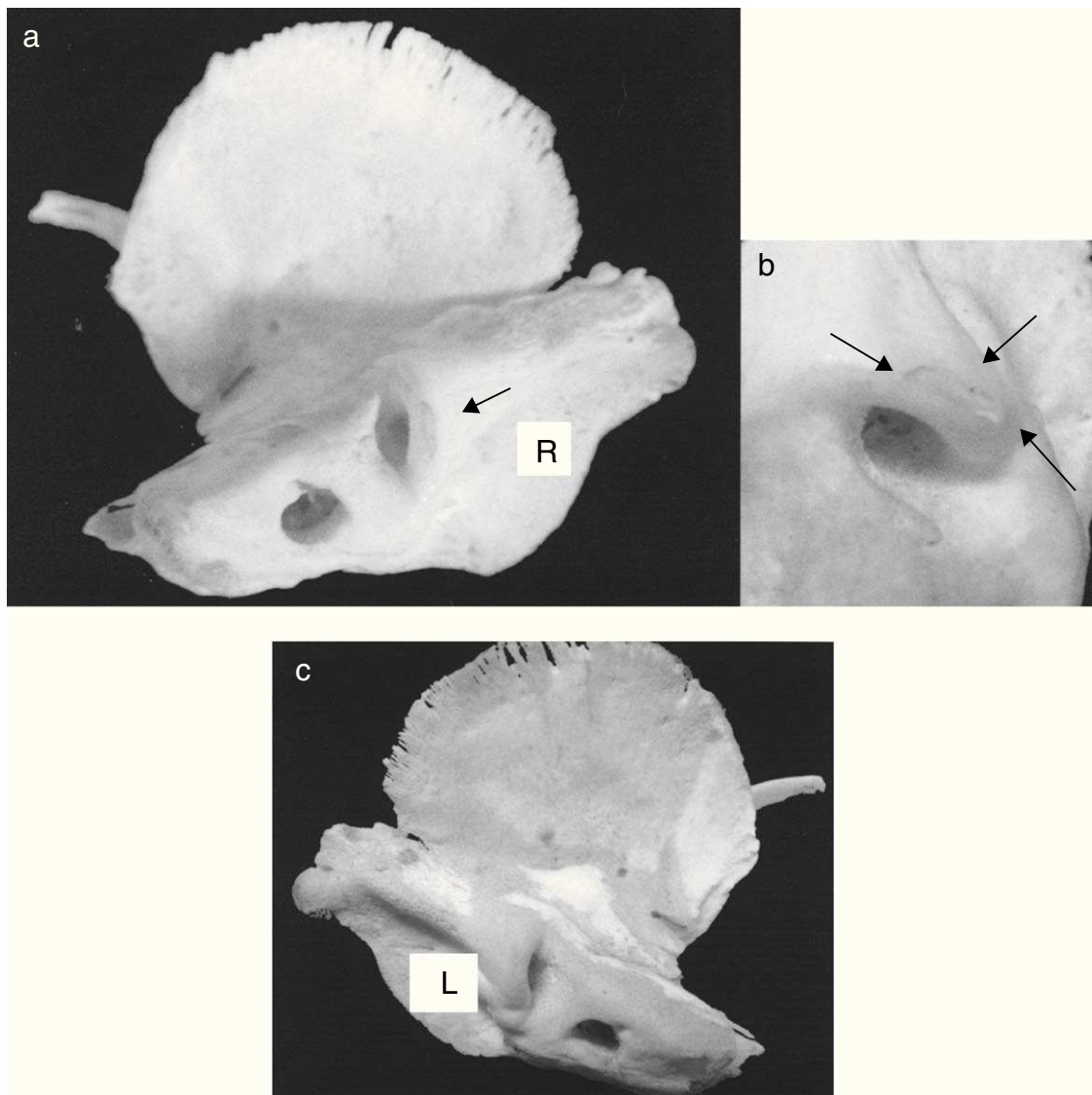
Fig. 1a shows the circumference of the superior semicircular canal and how it presents an elongated ellipsoid fossa (arrow) with smooth and soft edges. In one detail (Fig. 1b), we highlight how the canal exhibits a deterioration of its bony cover, formed exclusively by its internal or endosteal layer, which separates the canal from the superior semicircular duct.

Fig. 2a and b shows computed tomography (CT) scans of both temporal bones. Pöschl plane reconstructions of the superior semicircular canal showed that it was intact on both sides but we emphasise that bone coverage presented different thicknesses on the right side (a). The thickness also decreased along the curve of the canal (arrow) from 0.6 to 0.3 mm. On the left side, the thickness remained constant (arrows) around 0.5 mm.

## Discussion

The aetiopathogenesis of the anatomical change that causes the absence of bone covering in the superior semicircular canal is currently unknown, although several hypotheses have been postulated. Carey et al.<sup>2</sup> studied 1000 human temporal bones histologically. They suggested that the cause of the dehiscence might be an alteration of the postnatal development of the middle and outer layers of the 3 that form the cover of the canal. This author histologically demonstrated a uniform extreme thinness of the bone covering the superior semicircular canal in the middle cranial fossa at the time of birth, and its gradual thickening until 3 years of age. In addition, the thinning is frequently found bilaterally, and the pattern of ossification is generally stable, as evidenced by the presence of mature lamellar bone in the thinning margins, supporting this theory. In other words, it would be a change in the bone reorganisation of the otic capsule during the postnatal period.

Tsunoda and Terasaki<sup>3</sup> proposed that defects in the channel cover might have an embryological origin. To prove

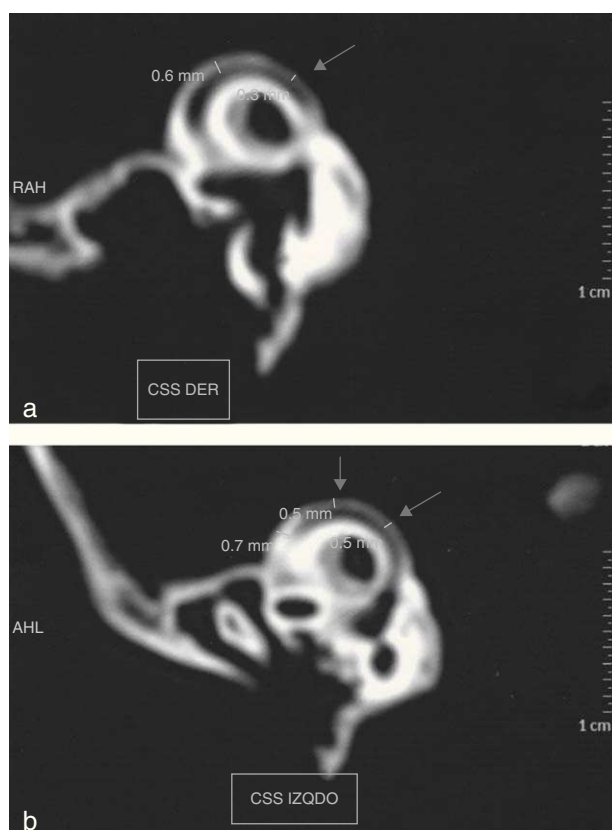


**Figure 1** Note in (a) and (b) an overview and detail of the right temporal bone, with alteration of the cover of the superior semicircular canal (arrow). We emphasise in (b) how the canal exhibits a deterioration of its bony cover, which is formed exclusively by its innermost or endosteal layer (arrows). Image (c) shows the left temporal bone, which is completely normal.

their theory, they used a computer simulation model that established that the cause of superior semicircular canal dehiscence is an incorrectly positioned primitive otocyst. When the otocyst is located very close to the developing brain, there is not enough space for the growth of the superior semicircular canal and the otic capsule comes into direct contact with the dura mater that originates mainly from the paraxial mesoderm. In this situation, the ossification of the inner and outer periosteal layers may not occur. The migratory pattern of loose reticular cells is altered and these mesenchymal cells are believed to be necessary for the completion of bone development of the labyrinthine structures. Consequently, the region may be left with a thin or incomplete development of the bone over the superior semicircular canal.

For Crovetto et al.,<sup>10</sup> the disturbance is prenatal and not postnatal. After studying 52 embryological foetal series from

6 mm to newborns, they found that in the development of superior semicircular canal, which begins its ossification at week 19 from 2 canalicular centres, there is a critical period of embryological development between weeks 24 and 28. In this critical period, there is a communication between the perilymph and the meningeal spaces of the middle cranial fossa through bone gaps. After this period, both spaces are separated by a thin, bony layer corresponding to the inner periosteum. In the foetus at 28 weeks, the semicircular canal is already surrounded by compact bone trabeculae, although the outermost layer of periosteum and the middle level are not fully developed, and it is still possible to observe bone gaps that maintain contact with the meningeal space. From week 30, the intermediate layer begins to disappear and the surface of the bone gaps becomes smaller, although the outer layer maintains contact with meningeal spaces in some sections. At 38 weeks, the 3 layers have



**Figure 2** Computed tomography and density study. Note how the superior semicircular canal was intact on both sides. We emphasise that, on the right side (a), reconstruction in the plane of the canal shows different widths of the bone cover, which decreases along the curve of the canal from 0.6 to 0.3 mm (arrow). On the left side (b), the width stays uniform, around 0.5 mm (arrows).

merged and the superior semicircular canal is surrounded by bone.

If this communication continues (due to a failure of the subsequent bone reorganisation of the 3 layers forming the cover of the canal) and its complete apposition does not take place, superior semicircular canal dehiscence will occur.

Brantberg et al.<sup>14</sup> and Mikulec et al.<sup>15</sup> believe that genetic factors or a congenital predisposition may be involved in the genesis of this ear disorder, as both have reported cases of dehiscence in siblings. Recently, Hildebrand et al.<sup>11</sup> have linked superior semicircular canal dehiscence with a mutation of the COCH gene.

After reviewing the CT scans of 131 temporal bones from children over 3 years, Chen et al.<sup>8</sup> observed 16 cases of superior semicircular canal dehiscence (8 on the right side and 8 on the left) in 14 patients (2 of them presented bilateral dehiscence). This presence in the paediatric population suggests that it may be the result of a developmental abnormality.

Hirvonen et al.<sup>12</sup> studied temporal bone CT scans comparing the thickness of the bone overlying the superior semicircular canal in patients with dehiscence syndrome (20 unilateral and 7 bilateral) with 88 control subjects. They showed that the thickness of the latter was greater

( $0.67 \pm 0.38$  mm) and correlated the thickness of one side with that of the other side. In addition, the width of the bone covering the superior semicircular canal in the intact side in patients with unilateral dehiscence was thinner ( $0.31 \pm 0.23$  mm) than in control subjects. These findings support the hypothesis of a developmental abnormality underlying superior semicircular canal dehiscence syndrome.

The frequent association of superior semicircular canal dehiscence with anatomical anomalies of the middle cranial fossa and middle ear also supports the hypothesis of a developmental abnormality. Gianoli<sup>16</sup> noted a high incidence of these anomalies, which included tegmen defects, geniculate ganglion dehiscence, temporal lobe encephalocele and cerebrospinal fluid fistula. Isaacson and Vrabec<sup>17</sup> support this increased incidence of dehiscent geniculate ganglion in patients with radiographic dehiscence and superior semicircular canal symptoms when compared with normal subjects.

Mahendran et al.<sup>18</sup> described a patient with a large tegmen defect in continuity with a superior semicircular canal dehiscence. This suggested a general deficiency in bone formation or bone deposition in the bottom of the middle cranial fossa or a dehiscent middle fossa with a common aetiology.

However, the theory that superior semicircular canal dehiscence is a developmental abnormality does not explain why some patients are asymptomatic and why the symptoms are not present in childhood, except for some rare exceptions.

To explain this, it has been hypothesised that in cases of an abnormally thin layer of bone covering the superior semicircular canal, a second event (such as a head trauma, a sudden increase in intracranial pressure or the erosion caused by the weight and pressure of the temporal lobe) may cause its complete rupture and dehiscence. This occurred in up to half of patients of Carey et al.<sup>2</sup> and was also a frequent finding for Minor,<sup>13</sup> who observed it in 23%, in a retrospective review of surgical and non-surgical patients with vestibular symptoms and superior semicircular canal dehiscence.

Watters et al.<sup>19</sup> described two women who presented symptoms and signs of acute superior semicircular canal dehiscence developed after delivery. They hypothesised that these patients had semicircular canals that were thin or minimally dehiscent (first event) and that the efforts against a closed glottis made during vaginal delivery caused an abrupt increase in intracranial pressure (second event); this in turn was responsible for the bone disruption of the posterior semicircular canals, with subsequent exposure of the membranous labyrinth, consistent with previous observations by Carey<sup>2</sup> and Minor.<sup>20</sup>

Ogutha et al.<sup>21</sup> cited another patient with imbalance, hearing loss and pulsatile tinnitus symptoms that appeared after a vaginal delivery. A CT scan showed that the patient suffered superior semicircular canal dehiscence. This was repaired surgically through the middle fossa and symptoms were resolved immediately.

It is also possible that this second event could destabilise the dura mater over a pre-existing dehiscence. Krombach et al.<sup>4</sup> argue that the increased flexibility of the dura mater covering the defect could be the event leading to the onset



of symptoms later in life. In this same line, Modugno et al.<sup>22</sup> believe that a dural tear could be the start of pathological manifestations after many years of silence of a likely malformation.

Carey et al.<sup>2</sup> did not find any arachnoid granulations eroding the bone overlying the superior semicircular canal in the temporal bones studied that showed thinning or superior semicircular canal dehiscence. Neither were other underlying bone diseases found to justify the loss of bone over the canal, except in one individual in whom the erosion observed was related to the presence of a vestibular schwannoma. This fact, together with the presence of bilateral cases, makes the alternative explanation that the bone is developed normally and is subsequently eroded symmetrically in later years less likely.

In the case described, we saw partial alteration of the cover of the superior semicircular canal on the right side, while the left was completely normal. The canal showed a deterioration of its bone cover formed exclusively by its innermost or endosteal layer, which separated the bone canal from the membranous superior semicircular canal, without formation of the middle and outer or periosteal layers. This event reflects a defect in canal development.

The CT and density study of both temporal bones showed how the superior semicircular canal was intact on both sides. However, we noted that the bone coverage on the right side presented a different thickness, which decreased along the curvature of the canal from 0.6 to 0.3 mm, while the left side maintained its thickness around 0.5 mm along the entire arc.

This would support the hypothesis that, in cases of an abnormally thin layer of bone covering the superior semicircular canal, a second event could cause its breakage and complete the dehiscence.

In general, when performing dissection of corpses, it is possible to observe how the dura mater is closely attached to the anterosuperior side of the petrosal and completely covers the canal. This leads us to believe that the defect would be covered by this membrane *in vivo*.

We believe that the dura mater must have an important role in the absence of symptoms in cases presenting dehiscence or defects in the canal, because it behaves like a true periosteum, closing the gap its bone coverage. This would explain why some patients are asymptomatic and why the symptoms are not present, with rare exceptions, in childhood, and why the symptoms are more common in older ages.

## Conclusion

The presence of partial defects in the bone covering the superior semicircular canal in the absence of its outer layers and middle layers, coupled with its smaller width, makes the canal susceptible to suffering a second event that may cause it to rupture and complete the dehiscence.

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