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Abnormal sella turcica morphology in individuals with congenital inner-ear hearing deficits

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Abstract

Introduction: *The present study focusses on the interrelationship between medically diagnosed congenital inner-ear hearing deficits and sella turcica morphology. Based on former prenatal studies it is hypothesized that the posterior wall and the floor of the sella turcica are malformed in patients with congenital inner-ear hearing deficits.*

Material and Methods:*The study is based on profile radiographs from 7 patients (10-17 years of age) with congenital inner-ear hearing deficits and hearing aids. The sella turcica was identified and described in these profile radiographs.*

Results: *The posterior walls and the floors of the sella turcicae are malformed in all 7 cases and the anterior wall appear close to normalin all cases. This observation proofs the involvement of the cranialoccipital field in congenital inner ear hearing deficits.*

Conclusion: *The observation of posterior wall and sella floor malformation observed on profile radiographs from patients with congenital inner-ear deficits has seemingly not been published before.*

In future analyses of profile radiographs, the morphology of the anterior wall, the posterior wall, and the floor of sella turcica should receive specific attention. Absence or malformation of the posterior wall could be a sign of congenital hearing deficit.

Keywords: *profile radiograph, inner-ear, craniofacial field, sella turcica, mastoid process*

Introduction

Since 1995 the prenatal morphology of the human sella turcica has been studied in human normal and pathological fetuses [1-16] These studies have demonstrated, that the anterior wall of the sella turcica has a neural crest origin and that malformation in this wall is associated with facial malformations [2,13-15]. Furthermore prenatal studies have revealed an association between the morphology of the posterior wall and the rostral end of the notochord [5,13]. Malformations in the posterior wall were found in malformed fetuses with spine- and cerebellar malformations [6,9,16]. The prenatal sella turcica morphology appeared different in different genotypes such as trisomy 21 and trisomy18 [6,8].

The floor of the sella turcica is the border area between the anterior and posterior part of the sella. This border area can be marked by a canal in the bottom of the floor stretching from the intra-cranial aspects to the extra-cranial aspects of the cranial base [13-15]. The presence of acanal could cause and explain the presence of a subpharyngeal location of the adeno-pituitary gland tissue, such as seen in severe facial malformation, including complete CLP patients [15,16].

Different cranial developmental fields have been localized to different areas of the sella. An overview of these regional areas or fields is demonstrated in figure 1. The different colors in this figure mark the fronto-nasal (yellow), the maxillary (red), the palatine (orange), neural crest fields [13-15]. Furthermore, the notochordal developed occipital field are highlighted in green in figure 1.

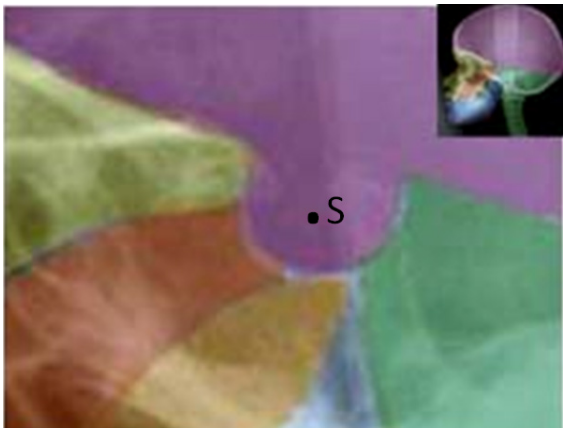

Figure 1. *Schematic drawing illustrating the shape of sella turcica with indication of the cephalometric landmark Sella, marked S. Anterior direction points to the left. The colouring of the walls and floor, which form the sella, refers to the different developmental fields: yellow = fronto-nasal neural crest field, red= maxillary neural crest field, orange = palatine neural crest field, green= notochordal developed occipital field. How the mandibular neural crest field (blue marking) is interrelated with the sella morphology has not yet been studied. The purple colour marks the theca field. Inserted in the figure is a profile radiograph, coloured according to the fields mentioned [14,15].*

The insight in the prenatal human sella turcica morphologies has in several articles been the background for the evaluation of the postnatal sella turcica morphologies [17-25].

The areas in the postnatal sella turcica reveals anterior wall, posterior wall and floor malformations with specific etiology [15,16]. Sella turcica malformations have also been associated with malformations inthe pituitary gland, the cerebrum, the cerebellum, and spine [2,13,15,18,19,23,25].

The present study focusses on the interrelationship between medically diagnosed inner-ear hearing deficits and sella turcica morphology. Prenatal studies have documented the location of the inner-ear in the mastoid process [13], demonstrated in figure 2. From an etiology point of view, the mastoid processes and the posterior wall of the sella turcica have developed both about 14 weeks of gestational age (GA), and both structures are formed by signaling from the notochord in the occipital field [15]. Accordingly it is hypothesized that there could be an association between malformation in the posterior wall and the floor of the sella turcica and congenital inner-ear hearing deficits in patients.

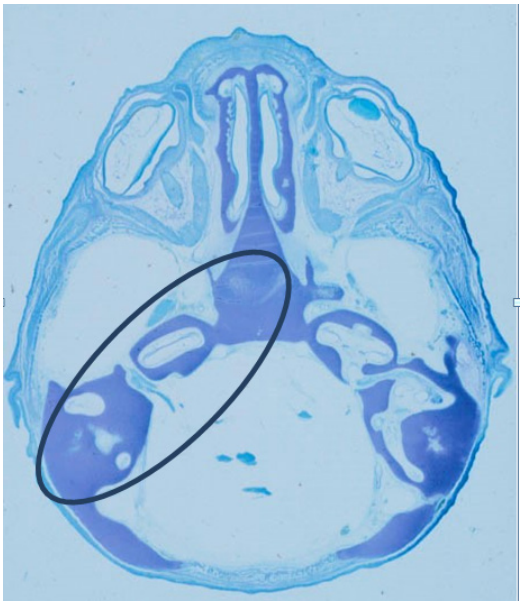

Figure 2. *Horisontal section of a cranial base from a human fetus, GA 14 weeks (13-15).Toluidine-blue [13-15].The area marked by a black oval conture marks the region of the mastoid process, and in the upper upper aspects of the oval conture is the cartilage forming the sella turcica located. Anterior points upward.*

Material and methods

During a period of 28 years profile radiographs, orthopantomograms and dental films have been forwarded from several countries to the author with questions concerning etiology-based diagnostics of dental and craniofacial deviations. Anamnestic information was sometimes available in the material referred and sometimes not. Information concerning medically diagnosed inner-ear hearing deficits and hearing aids, such as cochlear implants were available in 7 patients between 10 and 17 years of age (age unknown in one case). The present study is based on profile radiographs from these 7 patients. The sella turcica was identified and described from these radiographs.

Results

The radiographic appearances of the sella turcica in patients with congenital inner-ear hearing deficits are demonstrated in figure 3 A-H. In all figures, the anterior direction points to the left.

It is obvious from figure 3 that the malformations observed in the sella turcicae are different, ranging from minor malformations to more extended malformations. Also the involvement of the floor in these malformed sellae differs. Five cases (Figure 3 B,C,D,E and F) had complete or nearly complete absence of the posterior wall, and two cases demonstrated an abnormal bulky dorsum sella with a narrow floor and a caudal concavity or cleft in the inner aspect of the posterior wall (Figure 3 G,H).

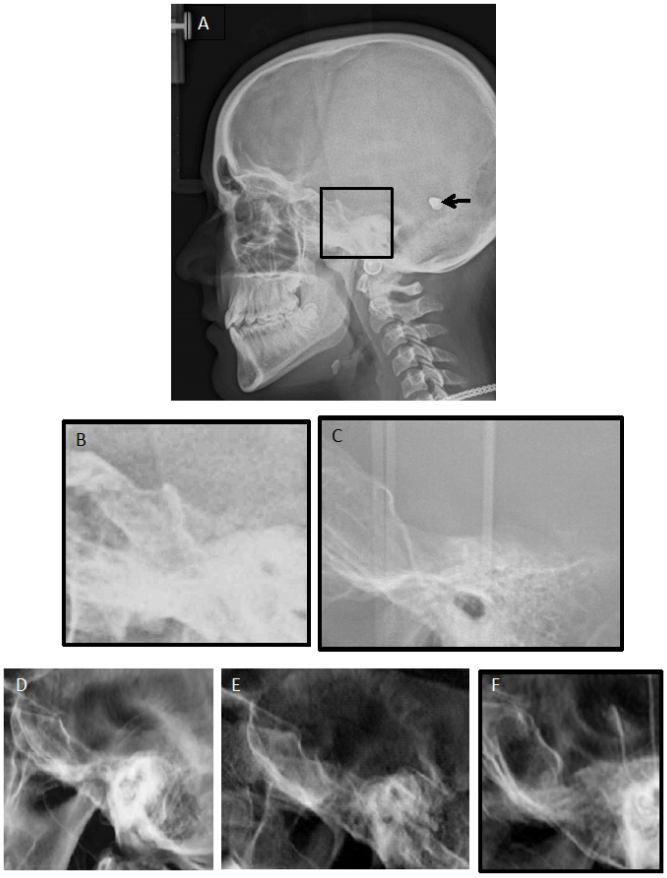

Figure 3. *These radiographic figures demonstrate the sella turcica area from patients with medically diagnosed hearing deficits. Anterior points to the left in all figures.*

A Profile radiograph from a boy (approximately 17 years old) with a quadratic marking of the sella turcica area. The arrow marks the cochlear implant.

B Enlargement of the marked area in fig. A. Note total absence of the posterior wall of the sella turcica.

C Radiograph, demonstrating the sella turcica area from a boy, 10 years of age. Note total absence of the posterior wall and a deepening of the floor in the sella turcica.

D Radiograph, demonstrating the sella turcica area from a girl, 14 years of age. Note total absence of the posterior wall of the sella turcica. Published with courtesy of Department of Dentistry and

Oral Health, Aarhus University, DK.

E Radiograph, demonstrating the sella turcica area from a girl, 12 years of age. Note total absence of the posterior wall of the sella turcica. Published with courtesy of Department of Dentistry and Oral Health, Aarhus University, DK.

F Radiograph, demonstrating the sella turcica area from a girl, 14 years of age. Note the absence of the posterior wall and a deepening of the floor in the sella turcica. Published with courtesy of Department of Dentistry and Oral Health, Aarhus University, DK.

G Radiograph of the sella turcica area from a boy, 11 years of age with hearing deficits. Note the abnormal developed posterior wall and a severe deepening of the floor of the sella turcica. Possibly a cavity can be observed in the inner aspect of the wall.

H Radiograph, demonstrating the sella turcica area from a boy, diagnosed with deafness (age unknown). Note the severely abnormal bulky posterior wall with a caudal concavity or cleft in the inner aspect of the wall.

It can be concluded that the posterior walls and the floors of the sella turcicae are malformed in all 7 cases with hearing deficits and that the anterior wall in all cases appear close to normal. This observation makes it possible to suggest the involvement of the occipital field (green area in Figure 1) in congenital inner ear hearing deficits.

Discussion

The seven patient profile radiographs in the present material represented all available profile radio-graphs from children and young adults with medically diagnosed hearing deficits, forwarded to the author during a period of 28 years.

The observation of posterior wall and sella floor malformations on profile radiographs from cases with congenital inner-ear deficits has previously been exemplified in the textbook "Etiology Based Dental and Craniofacial Diagnostics" [15], but seemingly not published scientifically based on an extended material before.

It can be hypothesized that the differences observed in sella turcica morphologies in the present study might reflect different malformations in the mastoid process. This aspect could be important to study in future studies in collaboration with otologists for clarifying if the specific nature of the hearing deficits reflects the different morphologies of the sella turcica.

In the cephalometric analyses the positioning of the S-landmark is important for the outcome of the orthodontic analyses (Figure 1). In the sella turcicae presented in this study (Figure. 3), the exact positions of the S-points areimpossible to define.

Axelsson et al. [26] described the variations in the normal morphology of sella turcica according to age. This study formed the basis for evaluating pathological morphologies of sella turcicae [26]. Interestingly, a bony bridging combining the anterior and posterior wall in the region of the diaphragm a sella has been observed in non-syndromic patients with severe craniofacial abnormalities [28].

The purpose of presenting the phenotypes of the sella turcica described in this article is to draw attention to the posterior sella wall on profile radiographs and specifically to the interrelationship between dorsum sella and the mastoid processes. In future orthodontic analyses description of the sella morphology should be included. The anterior wall, the posterior wall, and the floor should receive specific attention in this description.

Future studies should also focus on sella turcica phenotypes in different genotypes. A former study on sella turcica measured on profile radiographs fromgenetically identical twins demonstrated striking similarities in morphology between individuals within the same twin pair [29]. Also to the relationship between the sella and different categories of brain malformations are needed.

This present study focus on how sella turcica diagnostics on profile radiographs can supplement diagnostics by otologists. The sella turcica could be a key area for predicting not only deviations in craniofacial morphology [15,16], but also for the etiology-based diagnostics of the inner-ear abnormalities.

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