

Variations of The Isolated Cleft of The Hard Palate

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ABSTRACT

A retrospective study was undertaken of 3100 facial cleft cases in order to analyse and determine the incidence of the isolated cleft of the hard palate. This presented as a very rare occurrence in three different variations, each having specific clinical appearances.

The literature search revealed three publications each describing a single isolated hard palate cleft. However, these publications described hard palate clefts witnessed with other additional submucosal clefts or due to infection of the hard palate. They differ, in most cases, from the development and appearances of the clefts described in this paper.

This single paper describes 13 cases, demonstrating the three variations of the isolated cleft of the hard palate.

Key Words: Cleft palate, isolated hard palate cleft, cleft palate variation, median palatine cyst, oro-nasal fistula

INTRODUCTION

Cleft lip and palate deformities, or orofacial clefts, are generally classified according to their embryological development or appearance. The clefts are therefore classified as cleft lip, cleft lip and palate and cleft palate. However, this classification is not sufficiently detailed in relation to the clinical appearance - especially with reference to the various treatment options and techniques available for handling the orofacial cleft. For this reason, orofacial clefts seen at the University of Pretoria are classified according to the following types: cleft lip (CL), cleft lip and alveolus (CLA), cleft lip, alveolus and palate (CLAP), hard palate cleft (hP), hard and soft palate cleft (hPsP), and soft palate cleft (sP). Furthermore, detailed variations of these groups are also documented: for example, an isolated cleft lip (CL) with an isolated soft palate cleft (sP) would be recorded in detail as a combination cleft [COMBI], and an incomplete / partial soft palate cleft (sP) would be classified as a bifid uvula with a submucosal cleft¹.

The isolated cleft of the hard palate was recorded in 13 cases (0.42%) among 3100 cases of orofacial clefts at the University of Pretoria. These 13 cases could be subdivided into three distinct variations.

MATERIALS AND METHODS

The study population was drawn from the records of patients attending the outpatient Facial Cleft Deformity Clinic (FCDC) of the Department of Maxillo-Facial and Oral Surgery at the University of Pretoria. The database, extending over 24 years, was evaluated retrospectively. Orofacial cleft patients with an isolated cleft of the hard palate were selected for the study, based on the following criteria:

- No previous surgery.
- No cleft of the lip and alveolus.
- No cleft of the soft palate, including a submucous cleft.

The patients selected for this study were examined clinically at the time that they presented at the clinic and their particulars were carefully documented. The isolated clefts of the hard palate were classified and recorded according to a special cleft key,¹ which included the completeness (layers involved; cranial-caudal dimension) and the length (anterior-posterior dimension) of the cleft.

RESULTS

Among the 3100 patients suffering from oro-facial clefts, 1094 were recorded as cleft palate [hP (isolated hard palate), hPsP (hard and soft palate) and sP (isolated soft palate)], in the following ratios: 1,2 : 46,9 : 54,9 (hP : hPsP : sP). With an incidence of only 0,42%, the isolated cleft of the hard palate (hP) was extremely rare, only 13 cases being identified. Three different variations could be found: (1) a complete cleft, where all the layers are congenitally separated, total or partial in length; (2) an incomplete or submucosal cleft, where a lack of bony fusion promotes a blind soft-tissue fistula allowing food / foreign body impaction; (3) a second type of submucosal cleft, where the vomer-septum intercedes between the palatal shelves, thereby preventing the bony fusion of the palatal shelves.

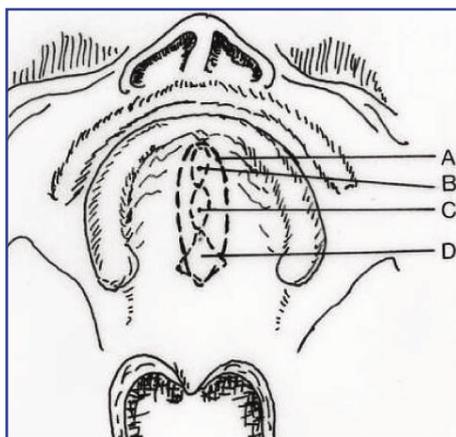


Figure 1a: Line drawing illustrating various clefts in the hard palate: A = total and complete isolated cleft of the hard palate; B = anterior partial isolated cleft of the hard palate; C = centrally located partial isolated cleft of the hard palate; D = posterior partial isolated cleft of the hard palate.



Figure 1b: A case with a total complete cleft of the isolated hard palate with a submucosal dento-alveolar cleft of the right side.



Figure 1c: A case with an anterior partial isolated cleft of the hard palate.



Figure 1d: A case with a centrally located partial isolated cleft of the hard palate.

THE THREE VARIATIONS OF THE ISOLATED HARD PALATE CLEFT AND THEIR FREQUENCIES

Total or Partial Complete Clefts

Six cases had presented with isolated clefts of the hard palate at birth. This is a complete cleft extending through all the tissue layers, i.e. no fusion had occurred. However, the length of the defect may vary, either extending antero-posteriorly over the entire hard palate (total) or occurring as discrete deficiencies in the mid-palatal area (partial) (Fig. 1a). One case presented with a total and complete cleft, i.e. full length of the hard palate and the separation of all layers (mucosal and bone). This patient also had a submucosal dento-alveolar cleft on the right side (Fig. 1b). Five cases had a partial complete isolated hard palate cleft, i.e. all layers were separated. However, these clefts were only partial in length and were situated somewhere between the incisive foramen and the posterior spine of the palatal bone. These five cases presented with differing anatomical locations of the cleft: one case had the cleft just posterior to the incisive foramen (Fig. 1c), three cases had the cleft in the central part of the hard palate (Fig. 1d), and in one case, the cleft was in the posterior hard palate and therefore at the junction of the hard and the soft palate (Fig. 1e). The latter cleft presented in a near-rhomboid form. The other clefts were ovoid in their appearances.

Incomplete clefts presenting as a sinus/cavity, which could in time result in a total breakdown (variation of a submucosal cleft)

The blind fistula or sinus may present as an incomplete cleft. As in the first group, separation of the oral mucosa and the bone occurs. However, the nasal mucosa is intact (Fig. 2a). Two patients presented with this type of cleft. One case had a sinus posterior to the incisive foramen and with entrapment of fingernail particles. The patient chewed his fingernails, and habitu-

ally had removed the impacted pieces from this cavity until the accumulation became too much for successful self-removal. During the surgical exploration and closure of this incomplete isolated cleft of the hard palate, 11 pieces of fingernail were removed. The second case, a healthy 56 year-old female, reported having been born with an intact hard palate yet, at the age of 26 years, developed a secondary complete isolated cleft of the hard palate. This cleft was relatively round in its appearance (Fig 2b).

Vomer-septum penetrations into the hard palate (second variation of submucosal cleft)

In this type of isolated hard palate cleft, the vomer bone-nasal septum penetrates through or intervenes between the hard palatal shelves in the oral cavity. During normal embryological development of the hard palate, the adjacent palatal shelves fuse thereby creating the midpalatal suture. The nasal septum (cartilage) and the vomer bone generally fuse onto the superior surface of the palatal shelves at the midpalatal suture (Fig. 3a). However, in these cleft cases the nasal septum-vomer complex is positioned between the palatal shelves, thereby maintaining separation of the palatal bone and resulting in the nasal septum-vomer complex penetrating into the oral cavity (Figs. 3b + 3c). In some cases, bony union is possible lateral to the nasal septum-vomer complex (see A in Fig. 3b), but mucosal fusion may also take place between the palatal shelves and this complex. The cleft may present as a submucosal isolated hard palate cleft (see B in Fig. 3b). Five patients were diagnosed with this cleft deformity of the hard palate (Fig 3d).

DISCUSSION

The embryological or developmental classification is based on the appearance of the two primordia of the primitive stomodeum, namely the primary palate - which gives rise to the lip and the alveolus with its posterior border at



Figure 1e: A case with a posterior partial isolated cleft of the hard palate

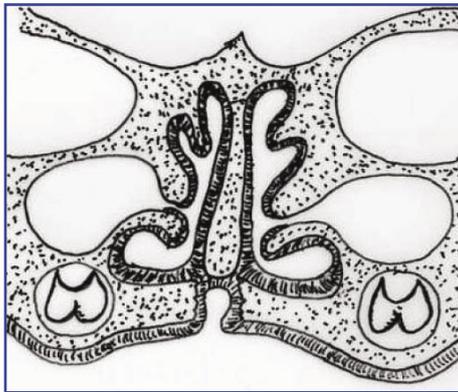


Figure 2a: Line drawing of an incomplete cleft, presenting as a sinus/cavity.

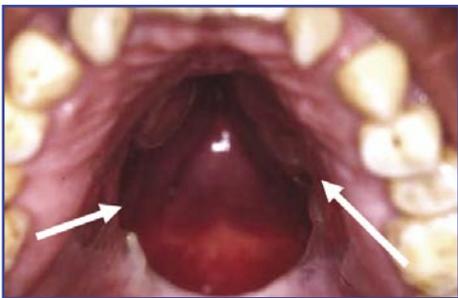


Figure 2b: A rounded type of isolated cleft of the hard palate, seen in a patient aged 56 years.

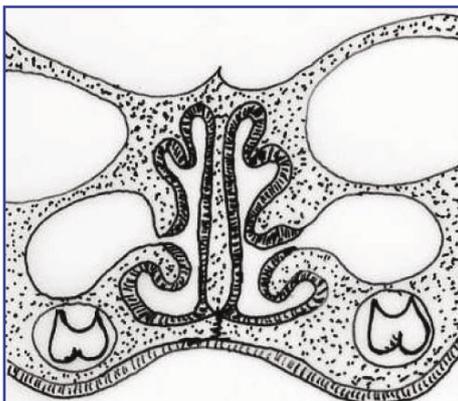


Figure 3a: A line drawing of the normal fusion occurring between the palatal shelves and the nasal septum.

the incisive foramen - and the secondary palate - which gives rise to the hard (bony) and soft palate. The fronto-nasal process forming the primary palate and the primitive nasal septum occasions the divisions of the stomodeal chamber into separate oral and nasal cavities. The secondary palate develops from the lateral palatal shelves, which are part of the maxillary processes. The tongue grows rapidly from the branchial arches, and quickly fills the oronasal cavity thereby forcing the lateral palatal shelves into an inferior position on either side of the tongue. Once the stomodeal chamber has enlarged and the head of the embryo becomes extended, the tongue moves downwards and the vertical palatal shelves are freed to swing horizontally towards each other and establish contact in the midline. It is generally believed that the nasal septum grows downwards to meet and then to fuse with the upper surfaces of the palatal processes after the palatal shelves have fused².

The term, isolated cleft palate, refers embryologically to a cleft of the secondary palate and is generally used for the hard and soft palate^{3,4}. The term isolated cleft of the hard palate therefore refers to the anterior part of the embryological secondary palate, and occurs between the bony palatal shelves. This type of cleft is very rare. The literature search revealed only three publications describing an isolated cleft of the hard palate^{5,6,7}. However, none of these publications referred to a true congenital isolated cleft of the hard palate, as all cases also involved submucosal clefts^{5,6} of the lip or soft palate, or occurred later in life spontaneously as a secondary isolated cleft of the hard palate^{6,7}.

One of these publications described the case of a painless swelling - formed a year before the patient had sought help - which had opened up into a spontaneous palatal fenestration and therefore a secondary isolated cleft of the hard palate. The authors assumed that this oronasal fistula had developed as a result of breakdown of the mucosa

covering an isolated cleft of the hard palate⁷. However, an infectious breakdown of a submucosal cleft or even of a median palatine cyst - with or without being embedded in a submucosal cleft - cannot be excluded, since the description of the case revealed the development of a swelling before the breakdown of the mucosal tissue. Furthermore, the photograph of the palate and dento-alveolar arches indicates an underdevelopment of the palate or partial collapse of the dento-alveolar arches, which may be explained by the presence of a submucosal cleft - as reported in a previous publication⁶.

The current study reports six cases with total or partial complete clefts. Only one of the six patients with a congenital isolated cleft of the hard palate presented with a total length of the hard palate (as well as with a concomitant submucosal cleft of the dento-alveolar arch)⁶. Five presented as partial complete isolated clefts of the hard palate since birth. Until now, five partial complete isolated clefts of the hard palate appear to be the only documented congenital isolated clefts of the hard palate in the literature^{5,6,7}.

As far as the incomplete or submucosal cleft (subsequently leading to sinus formation or cavity) is concerned, the complete enclosure of the palatal mucosa may possibly have led to the development of a fissural or developmental cyst known as a median palatine cyst^{8,9,10}. This cyst lies somewhere between the incisive foramen and the posterior spine of the hard palate and is not connected to the incisive foramen⁹. One case, a 56 year-old healthy female, presented with a complete round isolated cleft of the hard palate which occurred at the age of 26 years. One might presume that this case was originally an incomplete or submucosal cleft of the hard palate initially, or a median palatine cyst embedded in a submucosal cleft which, after breakdown of the cyst lining or until spontaneous perforation, resulted in this rounded type of isolated cleft of

the hard palate. This closely conforms to the description of a previously published case of an isolated cleft of the hard palate⁷. A complete cleft of the hard palate, as seen in bilateral CLAP and hPsP, is always ovoid and not round in its appearance.

An isolated cleft of the hard palate should not be confused with an oro-

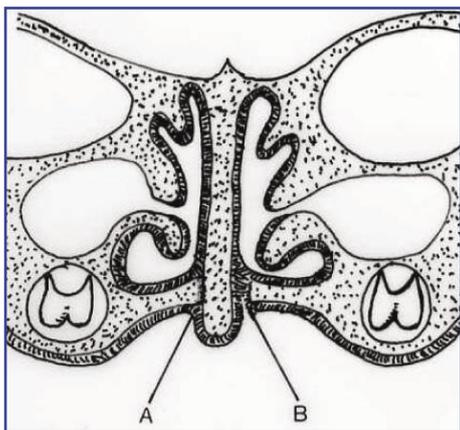


Figure 3b: A line drawing of a nasal septum-vomer penetration of the hard palate or intervention between the palatal shelves: A = bony fusion; B = mucosal fusion, as a submucosal cleft.

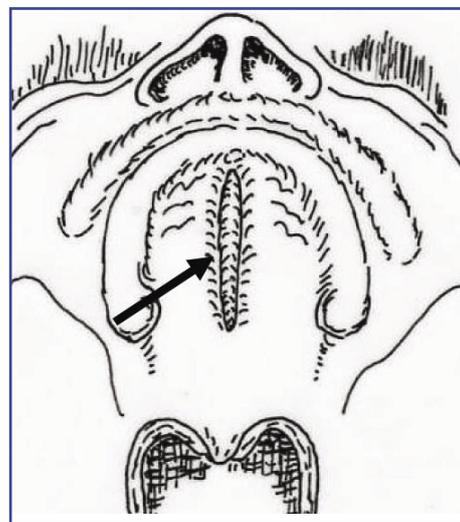


Figure 3c: A line drawing of an intra-oral view of nasal septum-vomer penetration of the hard palate



Figure 3d: A case with a nasal septum-vomer penetration of the hard palate.

nasal fistula, which occurs secondarily due to trauma,^{11,12,13} pathology¹⁴ (other than a possible median palatine cyst embedded in a submucosal cleft), or a resection or surgical complication^{11,15} of the bony part of the palate. Self-inflicted gunshot injury¹² to the submental region very often presents with a traumatic oronasal fistula in the hard palate. Certain pathological conditions give rise to a palatal oronasal fistula, such as Wegener's or other granulomatosis,¹⁶ tertiary syphilis, chronic dental infections and extraction¹⁷ with a discharging abscess (at the lateral part of the hard palate), breakdown of a benign¹³ or a malignant¹⁸ tumour, and others⁸.

SUMMARY

The FCDC of the University of Pretoria has a database extending over 24 years, comprising 3100 cases of cleft deformities. Thirteen (0,42%) of these cases were isolated clefts of the hard palate. Three variations of this cleft were observed: the total or partial complete isolated cleft of the hard palate; the incomplete or submucosal cleft of the hard palate which presents as a sinus, or becomes apparent after mucosal breakdown with or without a possible median palatine cyst; and the second variation of a submucosal cleft of the hard palate, where the nasal septum-vomer is positioned between the hard palate shelves, without bony fusion and which penetrates into the oral cavity.

CONCLUSION

Thirteen isolated clefts of the hard palate and their variations are documented and classified. The incidence of this type of cleft is extremely low.

Declaration: No conflict of interest was declared

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