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Original article

Is palato-premaxillary subduction a characteristic of Binder's syndrome?

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ABSTRACT

Objectives: Binder's syndrome is a rare malformative syndrome, defined clinically. CT study of a patient with this morphotype found palato-premaxillary joint subduction on a sagittal slice. The objective of the present study was to analyze this joint in a control population free of Binder's syndrome.
Material and methods: Fifty adult Caucasian patients who had undergone sinus CT scan between 2013 and 2016, showing normal nasolabial angle and with good visualization of the palato-premaxillary joint on a single sagittal slice, were selected. Joint analysis by 3 observers classified the patients in 2 groups: A, showing approximation between primary and secondary palate, and S, showing subduction. Alongside the observers' subjective analysis, the following parameters were compared: posterior palato-premaxillary angle (A_1), superior palato-premaxillary angle A_2, and the distance (d) of the premaxilla above the secondary palate.
Results: Fifty patients were included: 43 in group A and 7 in group S. A_1 angle differed significantly between groups: $111.95 \pm 10.22^\circ$ in group A, versus $130.53 \pm 10.0^\circ$ in group S; $P = 0.0015$. Values for A_2 and d did not differ according to group.
Conclusion: Two forms of palato-premaxillary joint, showing approximation or subduction, were found in the control population. Approximation was more frequent. Palato-premaxillary subduction does not in itself characterize Binder's syndrome.

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1. Introduction

Binder's syndrome is a very rare morphotype first described by Noyes in 1939 [1] and characterized by Binder in 1962 [2]. It is also known as maxillo-nasal dysplasia or maxillofacial dystosis. Binder first described it in 3 children with similar morphotypes, and defined several criteria: flat, vertical nose; nasal bone malpositioning; intermaxillary hypoplasia with malocclusion; hypoplasia or absence of anterior nasal spine; nasal mucosa atrophy; and (sometimes) absence of frontal sinuses. Several cases were subsequently reported, with wide clinical variations, and sometimes with associated deformities [3,4]. The other phenotypic criteria found in the literature comprise: acute nasolabial angle; short columella; horizontal half-moon nostrils; and wide philtrum [5,6]. All in all, the syndrome seems to be hard to define. One constant feature is the acute nasolabial angle: normal values range between 90° and 120°, for an esthetic "ideal" at 90–95° in males and 95–100° in females [7,8], whereas Binder's syndrome patients always show angles less than 70° (personal measurements from published photographs).

We treated a 19-year-old female patient corresponding to the Binder morphotype, for correction of nasal dysmorphia. Clinically, she presented with wide nasal dorsum, double nose tip, short bone dorsum, short columella, and asymmetric triangular nostril orifices. Laterally, she showed a very acute nasolabial angle, at 56°, with what seemed to be a very short columella and turned-up superior lip (Fig. 1). CT analysis found a particular aspect in the palato-premaxillary joint, showing subduction of the secondary palate under the premaxilla (primary palate) on medial sagittal slices (Fig. 2). The premaxilla looked to be tilted backward, overhanging the anterior extremity of the maxillary palatine apophyses, in contrast to the usual "approximating" aspect found between secondary palate and premaxilla (Fig. 3), where the palatine apophyses seem to abut the premaxilla, and the incisive canal seen to separate them on medial sagittal slices appears short and vertical.

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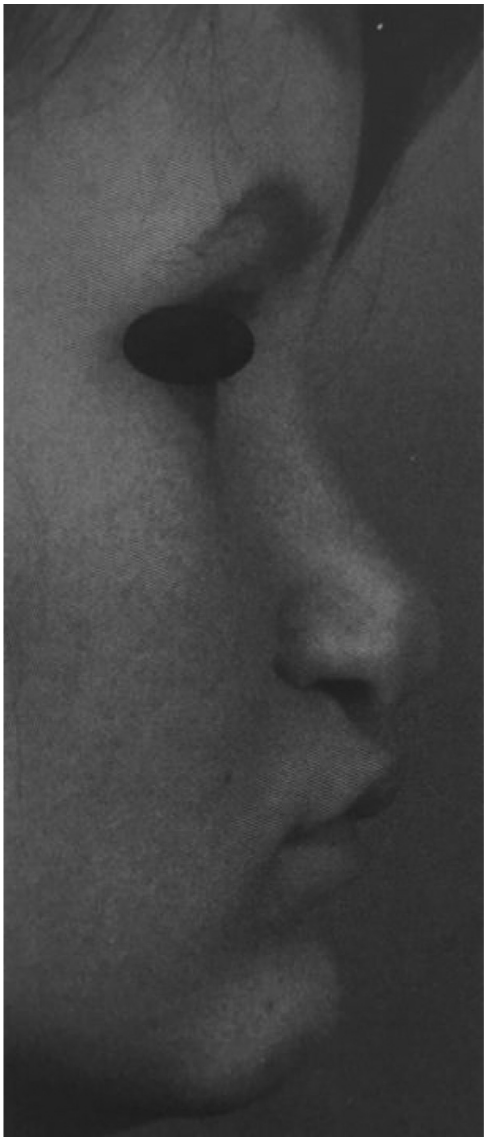

Fig. 1. Lateral photograph in Binder's syndrome.

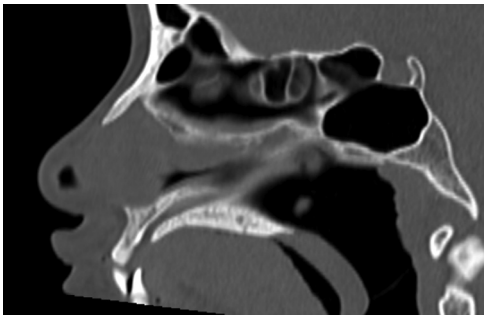

Fig. 2. Sagittal CT slice in Binder's syndrome, with velo-premaxillary subduction.

Embryologically, the secondary palate and respiratory nose develop via growth of the maxillary palatine plates [9]. These grow first downward on either side of the lateral edges of the tongue, then upward above the back of the tongue, quickly fusing together, anteriorly to posteriorly, along the mid-line. The anterior edges of the palatine plates anchor onto the posterior edge of the primary palate, leaving the incisive canal, which is a vestige of the primary choanae. The study hypothesis was therefore that, in case of palato-premaxillary subduction, palatine plate elevation and midline fusion are situated below the posterior edge of the premaxilla (primary palate), without approximation in the same plane, thus leading to premaxillary projection defect [10].

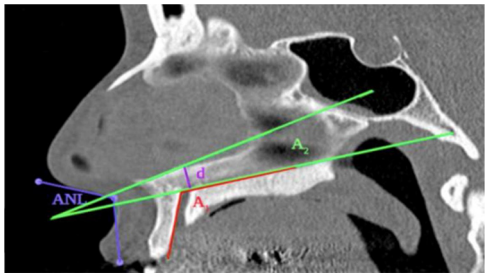

Fig. 3. Measurements in an "approximation" group subject. ANL: nasolabial angle; A_1: posterior palato-premaxillary angle; A_2: superior palato-premaxillary angle; d: distance of premaxilla above secondary palate.

The study objective was therefore to determine whether this aspect of palato-premaxillary subduction could be observed in a non-Binder population.

To test the hypothesis, the CT aspect of the palato-premaxillary joint was analyzed in a population presumed to be free of Binder's syndrome.

2. Materials and methods

A retrospective study included 50 adult caucasian subjects with sinus CT scan to assess chronic nasal dysfunction in the radiology department of our university hospital center between 2013 and 2016. Absence of Binder's syndrome was checked on finding nasolabial angle > 70° on CT (NLA: angle subtended by the columella and the superior segment of the upper lip). All subjects showed an anterior nasal spine and a straight nasal septum. The main inclusion criteria were good visualization of both incisive canal and velo-premaxillary joint on a single sagittal slice, and presence of an anterior nasal spine.

Exclusion criteria comprised history of maxillofacial or sinus surgery, and poor image quality (slice thickness > 3 mm).

CT scans were multiplanar in bone window. Mean slice thickness was 1.5 mm (range: 0.5–3 mm). CT used Aquilion One and Aquilion 64-slice scanners (Toshiba®) (native slices 0.5 mm and 1 mm, respectively) with 3D reconstruction on Osirix v.5.8.2 software for Mac.

Palato-premaxillary morphology was classified as either "approximation" between primary and secondary palates (group A, Fig. 3: the two structures face one another without overlap), or "subduction" of the secondary palate under the primary palate (premaxilla) (group S, Fig. 4). Group attribution was based on consensus between 3 observers (T.S.H, D.T.N, R.J), required to agree on presence or absence of subduction: i.e., aspect of secondary palate sliding under the premaxilla, as seen in our Binder's syndrome patient (Fig. 2).

On the assumption that these two joint forms would involve different angles around the joint, we performed the following measurements on each scan, on a sagittal slice through the incisive canal, on a millimeter ruler under Osirix software (Figs. 3 and 4):

- angle A_1: posterior palato-premaxillary angle subtended by the posterior edge of the premaxilla and the tangent to the anterior segment of the secondary palate (i.e., premaxillary tilt angle);
- angle A_2: superior palato-premaxillary angle subtended by the tangent to the anterior segment of the secondary palate and the tangent to the superior edge of the premaxilla;

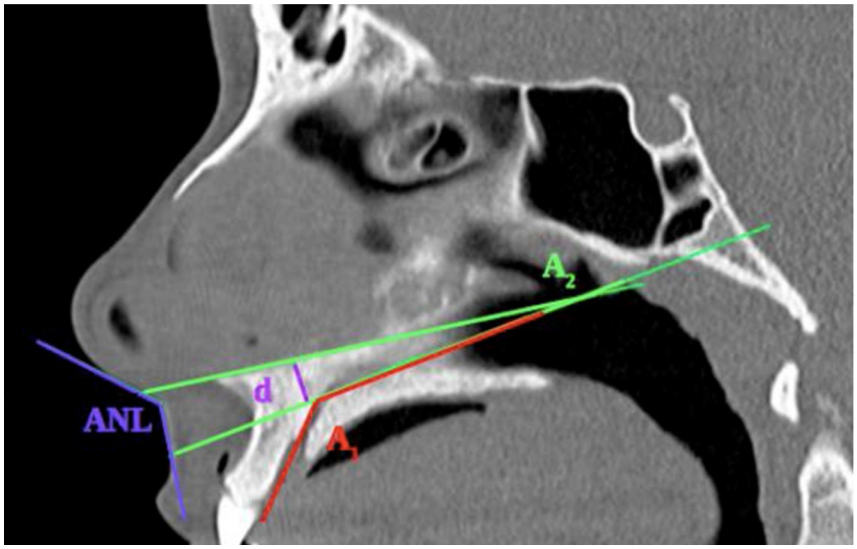

Fig. 4. Measurements in a "subduction" group subject. ANL: nasolabial angle; A_1: posterior palato-premaxillary angle; A_2: superior palato-premaxillary angle; d: distance of premaxilla above secondary palate.

Table 1
Mean values and standard deviations in groups A and S.

	Gr A ($n=43$)	Gr S ($n=7$)	P
Age (years)	39.36 ± 13.53	41.57 ± 19.91	0.85
F/M sex ratio	26/17	4/3	1.0
NLA	113 ± 15.5°	105 ± 19.8°	0.55
A_1	112 ± 10.2°	131 ± 10.0°	0.0015
A_2	15.3 ± 11.7°	9.5 ± 4.6°	0.36
d (mm)	4.3 ± 1.9	5.1 ± 2.1	0.3

A: approximation; S: subduction; F: female; M: male; NLA: nasolabial angle; A_1: posterior palato-premaxillary angle; A_2: superior palato-premaxillary angle; d: distance of premaxilla above secondary palate.

- distance d: premaxilla overhang above the anterior segment of the secondary palate, measured as the distance between the tangent to the anterior segment of the secondary palate and the highest point of the palato-premaxillary joint.

Statistical analysis used SAS 9.1 software (SAS Institute, Inc., Cary, NC). Inter-group comparison used Fisher exact and Wilcoxon–Mann–Whitney tests for continuous variables. The significance threshold was set at $P < 0.05$.

3. Results

Fifty CT scans were included: 20 male and 30 female subjects. Forty-three patients (17 male, 26 female; mean age, 39.4 ± 13.5 years) showed palato-premaxillary approximation (group A), and 7 (3 male, 4 female; mean age, 41.6 ± 19.9 years) showed subduction (group S). The two groups were comparable for age ($P = 0.85$) and gender ($P = 1$).

Mean nasolabial angle (NLA) was 112.8 ± 15.5° in group A and 105.3 ± 19.8° in group S ($P = 0.55$).

Mean A_1 angle differed significantly between groups: 112 ± 10.2° in group A versus 130.5 ± 10.0° in group S; $P = 0.0015$. Mean A_2 angle did not differ significantly: 15.3 ± 11.7° in group A and 9.5 ± 4.6° in group S ($P = 0.36$).

Mean distance d was 4.3 ± 1.9 mm in group A and 5.1 ± 2.1 mm in group S ($P = 0.3$) (Table 1).

The Binder's syndrome patient showed values of: NLA = 56°; A_1 = 140.2°; A_2 = 8.9°; and d = 4.9 mm (Table 2).

In both groups, parameters did not differ significantly according to gender (Table 3).

Table 2
Mean values in Binder's syndrome patient.

Age (years)	19
Gender	F
NLA	56°
A_1	140.2°
A_2	8.9°
d (mm)	4.9

F: female; NLA: nasolabial angle; A_1: posterior palato-premaxillary angle; A_2: superior velo-premaxillary angle; d: distance of premaxilla above secondary palate.

4. Discussion

Finding palato-premaxillary subduction in the present case of Binder's syndrome led us to study this joint in a non-Binder control population. Normal nasal morphology is defined subjectively in terms of harmony and esthetics, and we sought a simple criterion characterizing Binder's syndrome, such as the CT aspect of the joint. In the literature, nasolabial angle is systematically acute in Binder's syndrome, never exceeding 70° in published reports, when specified [11]. We therefore arbitrarily selected the non-Binder population among patients with NLA > 70°. Measurement confirmed that it was much more obtuse in controls, at a mean 112° taking males and females together, than in the Binder patient (56°).

Subjective classification according to palato-premaxillary approximation or subduction found a majority (86%) for approximation, with only 7 patients (14%) showing subduction. Age, gender and NLA were comparable between groups. A_1 angle (i.e., incisive canal inclination), on the other hand, showed a significant difference, with greater obtuseness correlating with subduction; comparably, the Binder patient showed an A_1 angle of 140.2°. Subduction of the secondary under the primary palate would seem to cause the joint to slide, and logically makes A_1 more obtuse. In approximation, the secondary palate pushes against the primary palate, verticalizing the incisive canal, resulting in a more acute A_1 angle.

Measurement of A_2 angle and distance d sought to identify a premaxillary projection defect due to the palatine apophyses not pushing against the premaxilla. Distance d measured premaxillary overhang above the secondary palate. The two groups did not significantly differ on these parameters. This may have been due the difference in group size, with preponderance of approximation, and

Table 3
Mean values and standard deviations according to group and gender.

	Gr A ($n=43$)			Gr S ($n=7$)		
	Female ($n=26$)	Male ($n=17$)	P	Female ($n=4$)	Male ($n=3$)	P
Age	41.38 ± 12.98	36.18 ± 14.14	0.22	43.25 ± 20.9	39.33 ± 22.81	0.86
NLA	114.42 ± 12.68	110.39 ± 19.18	0.57	98.15 ± 24.32	114.9 ± 6.95	0.41
A_1	113.54 ± 9.7	109.52 ± 10.82	0.21	136.33 ± 4.82	122.80 ± 10.4	0.26
A_2	12.41 ± 8.82	19.72 ± 14.25	0.099	10.28 ± 4.91	8.57 ± 4.84	0.86
d (mm)	4.11 ± 2.08	4.63 ± 1.65	0.23	5.73 ± 2.18	4.23 ± 1.95	0.41

A: approximation; S: subduction; NLA: nasolabial angle; A_1: posterior palato-premaxillary angle; A_2: superior palato-premaxillary angle; d: distance of premaxilla above secondary palate.

thus lack of power. There was nevertheless a trend toward a more acute A_2 angle in group S. When the secondary palate does not approximate the primary palate, the lack of pressure between the two may make the superior palato-premaxillary angle A_2 more "parallel", becoming acute or even non-existent in some cases. However, no conclusion can be drawn on this point, given the wide variation found within group A, from 0° to 46°. Moreover, depending on the curvature between the proximal and distal parts of the secondary palate, A_2 angle may be inverted within the same group, without this being related to the absolute value of the angle itself.

Distance d (premaxillary overhang above the anterior part of the secondary palate) showed no significant difference between groups. Premaxillary elevation over the anterior part of the secondary palate was similar, regardless of the form of the joint. When the secondary palate closes below the premaxilla during embryological development, there may still be some impaction force pushing the premaxilla upward. The main defect may be one of projection forward rather than upward; distance d does not measure this.

NLA was slightly more acute in group S (105°) than group A (113°), although not significantly ($P=0.55$). The group size difference precluded any definite conclusion, but subduction of the secondary under the primary palate may contribute to NLA closure. Backward tilt of the premaxilla may induce invagination of the superior lip under the columella, producing an acute NLA, although other factors may also be relevant, such as tegument trophicity, facial muscle implantation and cartilage growth.

According to evo-devo theory, respiratory septum formation by back-to-back verticalization of the vomers takes place upward and backward by impaction in the incisive canal [12]. Vomer growth progresses between the fused secondary palate below, the olfactory septum above, the sphenoid behind and the primary palate in front. In our Binder patient, palatine plate fusion thus took palace below rather than behind the primary palate, which tilted backward; the hypothesis of an embryological origin of Binder's syndrome was borne out in a prenatal diagnostic study with 2 cases [13]. Some authors also suggested that vitamin K deficit during pregnancy could be implicated [14].

Although subduction of the secondary under the primary palate was also seen, approximation was clearly predominant in the non-Binder population. While subduction cannot be considered specifically pathognomic for Binder's syndrome, it may still be a necessary variant, the pathological phenotypic consequences resulting from association with other factors.

Given the phenotypic variability and subjective clinical criteria in Binder's syndrome, several radiological parameters would be interesting to identify. Lateral radio-cephalometry confirms that the skeletal abnormalities are mainly located in the maxilla, with mid-face hypoplasia in the vertical and anteroposterior directions [15]. 3D cephalometric studies would also be interesting, to demonstrate global maxillary hypoplasia and analyze other radiological criteria [16], such as orbital edge retraction and dental arcade disorder. Multi-ethnic studies could compare velo-premaxillary aspect in Asian and African subjects. These findings would be worth confirming by studying several cases of Binder's syndrome; but these are rare, and those with CT scan even more so. We sought in vain for CT files in other French university hospitals. A prospective study including however few cases has been considered, as better radiologic knowledge of the syndrome could help diagnosis, and also enable treatment to be adapted to our understanding of the deformity [17–19].

5. Conclusion

Two forms of palato-premaxillary joint can be found in the general population, with either subduction or approximation between the primary and secondary palates. Subduction is not pathognomic for Binder's syndrome, as several cases were observed radiologically in subjects with normal nasolabial angle. However, the present study found that subduction was rare in the control population.

Disclosure of interest

The authors declare that they have no competing interest.

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