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CLINICAL SECTION

Maxillo-nasal dysplasia, Binder's syndrome: review of the literature and case report

F. M. V. Dyer
D. R. Willmot
Charles Clifford Dental Hospital, UK

Abstract

A 12-year-old girl with maxillo-nasal dysplasia (Binder's syndrome), featuring maxillary hypoplasia and relative mandibular prognathism, presented with a Class III incisal relationship. Her treatment was managed orthodontically. The principal features of the syndrome and management of these cases is discussed.

Key words:
maxillo-nasal dysplasia.

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Introduction

Despite the frequent presence of dental malocclusion in patients with maxillo-nasal dysplasia, very little is to be found in the orthodontic literature. Traditionally, plastic surgeons have been more closely involved with the syndrome, presenting different methods of surgical correction to solve the aesthetic and/or functional problems.[1–3]

The essential features of maxillo-nasal dysplasia were initially described by Noyes in 1939,[4] although it was Binder who first defined it as a distinct clinical syndrome. He reported on three cases and recorded six specific characteristics:[5]

- Arhinoid face.
- Abnormal position of nasal bones.
- Inter-maxillary hypoplasia with associated malocclusion.
- Reduced or absent anterior nasal spine.
- Atrophy of nasal mucosa.
- Absence of frontal sinus (not obligatory).

Individuals with Binder's syndrome have a characteristic appearance that is easily recognizable.[6] The mid-face profile is hypoplastic, the nose is flattened, the upper lip is convex with a broad philtrum, the nostrils are typically crescent or semi-lunar in shape due to the short collumela, and a deep fold or fossa occurs between the upper lip and the nose, resulting in an acute naso-labial angle. Cephalometrically, there is a reduced sella–nasion distance[7] and the length of the maxilla measured from the anterior surface to the posterior nasal spine is reduced, partly due to the mid-face skeletal defect that extends from the inferior rim of the piriform aperture beyond the apical base.[2]

Maxillo-nasal dysplasia can also be combined with other malformations. For example, Olow-Nordenram and Radberg reported 44.2 per cent of a study sample to have malformation of cervical vertebrae.[8] The association with pseudo-mandibular prognathism has also been described.[2, 5, 9] In the most severe cases, the syndrome is associated with true mandibular prognathism, which requires combined orthodontic and surgical treatment.[10]

Aetiology

When we consider the aetiology of this condition Binder suggested that there was a disturbance of the prosencephalic induction centre during embryonic growth.[2] However, it has been suggested that there is a common concurrent induction process for both the prosencephalic area and the vertebrae, accounting for the increase of vertebral anomalies associated with the condition.[8] Birth trauma has also been suggested as a possible causative factor, but is not further substantiated in the literature.[4] The possibility of a family history was put forward by Ferguson and Thompson.[11] However, Olow-Nordenram and Valentin were unable to disprove the possibility of a genetic aetiology in a study of 50 patients with the condition, involving 60 families.[12] In a further study of 97 individuals with

Address for correspondence: Dr F. M. V. Dyer, Orthodontic Department, Charles Clifford Dental Hospital, Wellesley Road, Sheffield S10 2SZ, UK.

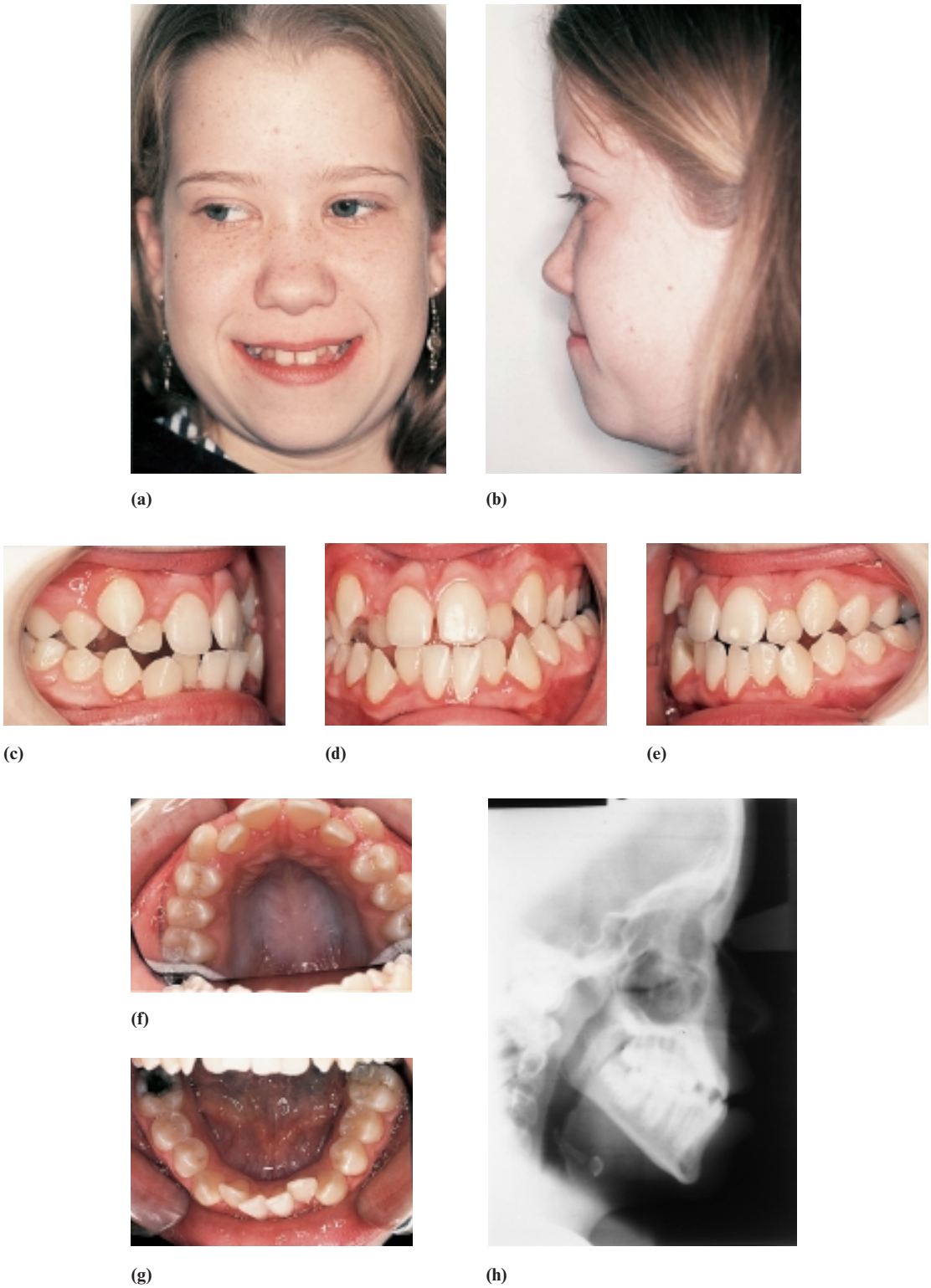


Fig. 1 (a–h) Case report: pre-treatment records.

Binder's syndrome, Olow-Nordenram[13] reported a positive family history was for 36 per cent.

Gorlin *et al*. suggest that maxillo-nasal dysplasia is a non-specific abnormality of the nasomaxillary complex. They believed that familial examples are a result of complex genetic factors, similar to those involved in producing a malocclusion.[14]

It is generally agreed that the lack of population frequency data has affected the evaluation of aetiological findings.

Dental features, diagnosis, and treatment planning

Holmstrom reported that 5 per cent of his Binder's cases presented with Class III malocclusions. There may be pseudo-mandibular prognathism or true mandibular prognathism combined with a hypoplastic maxilla.[2]

The severity of the malocclusion is ultimately connected with the severity of the syndrome. In mild cases, orthodontic treatment may not be necessary because of compensatory effects in the dental arches, while in the most severe cases the maxillary under-development is aggravated by the mandibular prognathism, and can only be treated with a combination of orthodontics and surgery.

Case reports have described morphological characteristics of the syndrome, which are of fundamental importance for correct diagnosis and treatment planning. The following measurements are shorter or smaller than normal: anterior cranial base length, cranial base angle, inclination of the nasal bones, upper apical base angle, maxillary length, upper anterior face height, pharyngeal depth, facial convexity, and nasal prominence. There was also wide variation in measurements with age, the mandible becoming more prognathic with a corresponding increase in the gonial angle. In addition, angular measurements, such as the mandibular planes angle and nasal plane (ANS–PNS) to mandibular planes angle, were found to be significantly increased.[10]

Furthermore, Munro reported that eight of 11 patients in his study had an increase in mandibular length.[1] In addition, a longitudinal study of 13 untreated cases reported a significantly shorter mandibular ramus length when compared to a control group.[10] In addition, proclination of the upper incisors was a significant finding as they compensate for a short and retrognathic maxilla, thereby facilitating an acceptable occlusion. These authors also reported a wide variation of lower incisor inclination, suggesting no trend specific to maxillo-nasal dysplasia.

Case report

A 12-year 11-month-old female, diagnosed with maxillo-nasal dysplasia, attended the orthodontic department following referral from a consultant plastic surgeon. The patient requested straightening of her teeth. Eighteen months previously she had failed a course of functional appliance therapy due to poor compliance after only 3 months of wear.

On examination she presented with a Class III incisal relationship on a mild Class III skeletal pattern. The Frankfort mandibular planes angle and the lower face height were increased. Mid-face hypoplasia was evident with an absence of fronto-nasal angle reflected in a straight profile. There was also reduced sagittal development of the nose. Transversally there was no apparent facial asymmetry. The lips were competent at rest.

Intra-oral examination of the dentition revealed the presence of all permanent teeth apart from the third molars. There were occlusal restorations in LL6 and LR6. The oral hygiene was of a poor standard and there was generalized marginal gingivitis.

There was moderate crowding of the lower arch with the lower incisors being retroclined. The upper arch was moderately crowded with the lateral incisors palatally placed. The overjet and the overbite were edge-to-edge in a retruded contact position. The upper centreline was coincident with the face and the lower displaced to the left by 3 mm. There was no displacement or deviation on opening or closing. The molar relationship was a full unit Class III on the right and left (Figure 1a–h).

The panoramic radiograph showed the presence of all third molars. There was no evidence of caries.

Standard cephalometrics are not appropriate in Binder's patients because the hypoplastic anterior nasal spine combined with thinness of labial plate of alveolar bone over the upper incisors make the position of point A difficult to determine. An alternative cephalometric analysis has been suggested and used by Olow-Nordenram and Thilander following Bjork's analysis (Figure 2).

The lateral cephalogram in this patient indicated a Class III skeletal pattern using Olow-Nordenram analysis (Table 1). The maxilla was retrognathic and mandible demonstrated pseudo-mandibular prognathism. The maxillary mandibular planes angle was increased at 38 degrees, however the lower anterior face

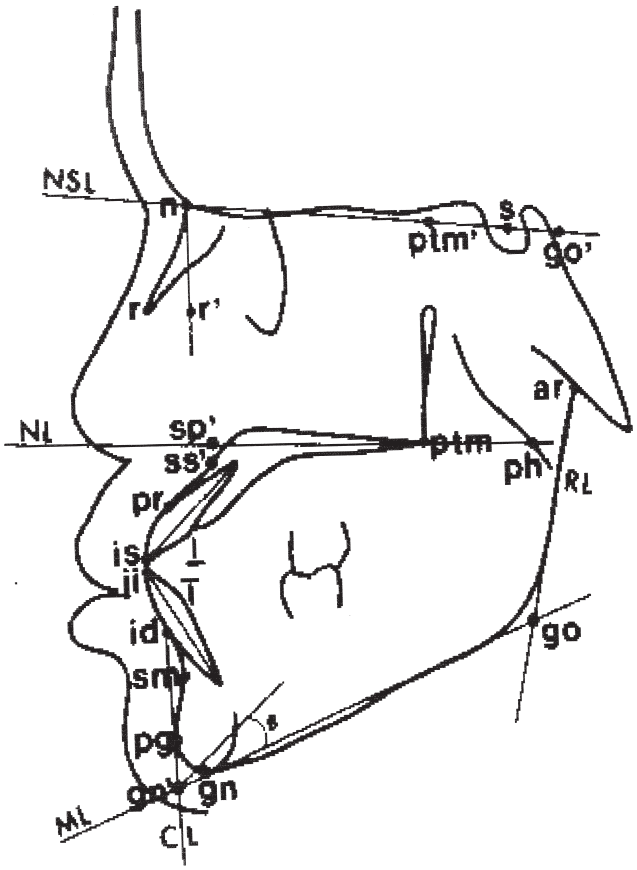


Fig. 2 Special points and lines used in Olow-Norderam (1987),[15] modification of those used by Bjork. CL = chin line, the tangent to the chin through id (gn′–id); gn′ = intersection between the chin line (CL) and the mandibular line (ML); go′ = projection of gonion on the nasion–sella line (NSL); ptm′ = projection of ptm on NSL; ss′ = deepest point on the anterior contour of the upper alveolar process at the level of the apices of the central incisors; sp′ = projection of ss′ on the nasal plane (NL); NL = midline of the palate through ptm; ph = intersection between NL and the contour of the posterior pharyngeal wall; and r = rhinion, the most antero-inferior point on the nasal bone.

height was 55 per cent of the total anterior face height. Dentally, the upper incisors were proclined to the maxillary plane at 114 degrees and the lower incisors retroclined at 85 degrees, compensating for the skeletal pattern and the increased maxillary mandibular planes angle.

The aims of treatment were:

- relieve crowding;
- level and align upper and lower arches;
- obtain positive overjet and overbite;
- correct the lower centreline;
- co-ordinate the arches;
- obtain a Class I molar relationship.

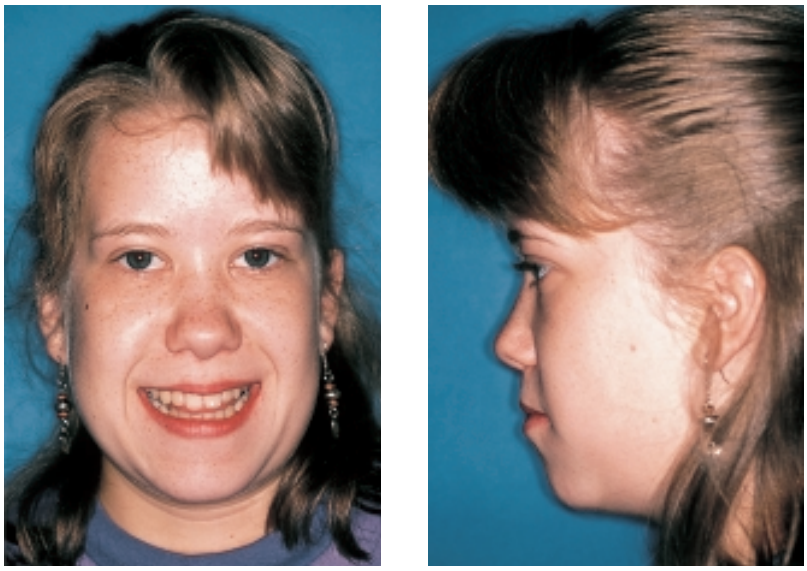
Table 1 Case presentation: pre- and post-treatment cephalometric analysis using Olow-Nordenram modification of Bjork's analysis

	Pre-treatment	Post-treatment
Cranial		
n–s (mm)	61	63
n–s–ar (°)	110	129
Facial upper		
s–n–ss′ (°)	77	77
SN/max plane (°)	12	15
n–sp′ (upper anterior face height) mm	54	59
Sp′–ptm′ (maxillary length) mm	42	44
Ptm–ptm′ (upper posterior face height) mm	45	49
Facial lower		
s–n–b (°)	77	76
Ar–go–gn (gonial angle)	134	136
NSL–ML (°)	50	52
Sp′–gn (lower anterior face height) mm	68	73
Go–ar (ramus length) mm	37	39
Gn–go (mandibular body) mm	72	78
Facial upper & lower		
Max-mand planes angle (°)	38	36
s–ar–go (articular angle, °)	148	148
n–gn (anterior face height) mm	122	134
Go–go′ (posterior face height) mm	65	68
Pharyngeal depth (mm)	20	18
Dental relationships		
Upper incisor/max plane	114	115
Lower incisors/mand plane	85	72
Inter-incisal angle	125	136

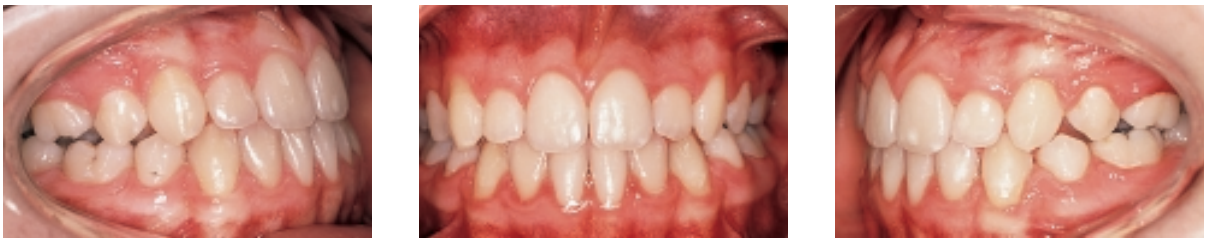
The treatment plan was a follows:

- improve oral hygiene;
- extraction of UR5 UL5 LL4 LR4;
- upper and lower pre-adjusted Edgewise appliance using Andrew's prescription brackets (0.022 × 0.028-inch slot size).

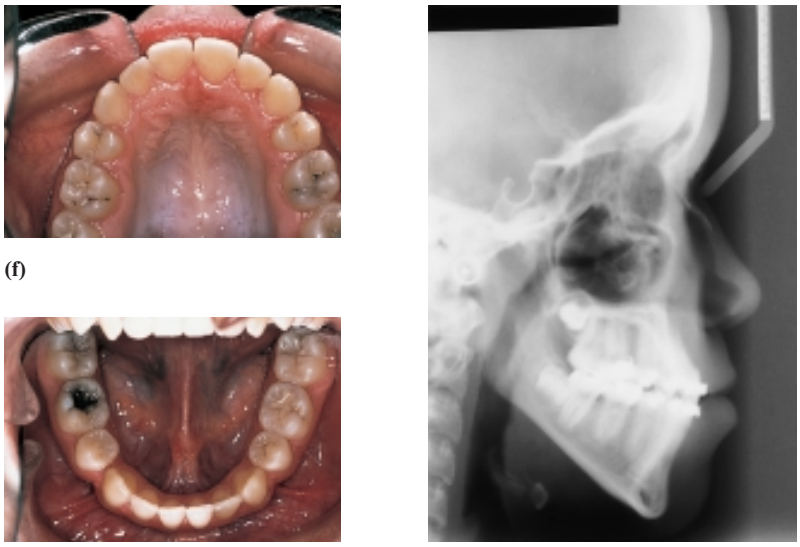
Treatment was completed in 19 months. Working archwires of an upper 0.019 × 0.025-inch rectangular SS and lower 0.018-inch SS round wire with circle hooks between the lower lateral incisors and canines, enabled space closure and facilitated dentoalveolar camouflage. Light Class III elastics were used for 4 months towards the end of treatment. Following debond, upper and lower Hawley retainers were provided. These were worn full time for 8 months and a further 10 months at night only (Figure 3a–h).



(a) (b)



(c) (d) (e)



(f) (g) (h)

Fig. 3 (a–h) Case report: post treatment records.

Case assessment

This patient with maxillo-nasal dysplasia presented with a Class III incisal relationship on a mild Class III skeletal base. Asymmetric premolar extractions were used between the arches to allow differential movement of the molars for correction of the Class III buccal segment relationship. The Class III incisal relationship has been managed by camouflage. The position of the upper incisors has been maintained and the lower incisors have been further retroclined during treatment.

Cephalometric superimposition demonstrates an opening of the cranial base angle (n–s–ar), which has displaced the facial complex vertically. There has been minimal change in maxillary length and the increase in mandibular length has contributed to the posterior rotation (Figure 4).

The stability of this case depends on the potential for any further mandibular growth. An adequate overbite should aid retention of the overjet correction and this has remained stable 24 months post-debond.

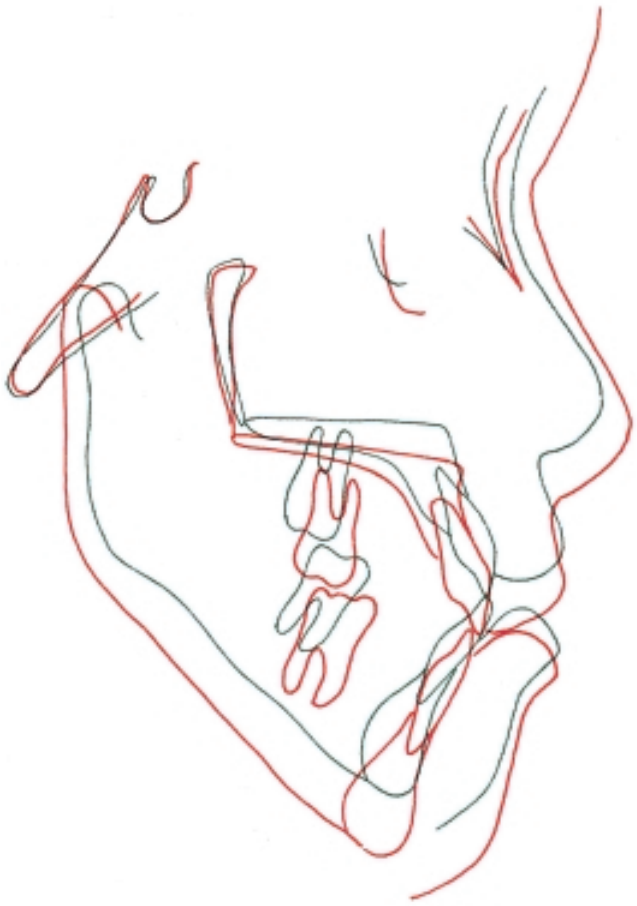


Fig. 4 Superimposition of pre-treatment and post-treatment cephalogram.

Maxillo-nasal dysplasia and long-term growth potential

In longitudinal cephalometric studies of children with Binder's syndrome, comparing orthodontically treated cases with untreated cases, it was concluded that conventional orthodontic therapy did not produce evidence for a positive influence on craniofacial growth.[10] With increasing age the maxilla grew forward, but not to the same extent as the mandible. Growth impediment was confined to the area around the absent anterior nasal spine, in subjects with moderate forms of the syndrome.

The mandible grew in length, in the body and ramus. The initial smaller length of the mandible seen between the ages 9 and 14 years was not evident later when compared to controls suggesting that 'catch up' growth of the mandible occurred after this age. Generally, a posterior rotation of the mandible was seen with growth.

Orthodontic treatment planning in maxillo-nasal dysplasia patient

Olow-Norderam and Thilander advised postponing definitive orthodontic treatment in individuals with maxillo-nasal dysplasia until growth has stopped, especially in those with a severe malocclusion.[10] It has been suggested that corrective surgery of the mid-face and nose has the potential to jeopardize acceptable occlusal results following early orthodontic correction.

Olow-Norderam concluded that the severity of the malocclusion was evident at an early age. Patients who proceeded on to orthognathic surgical correction had more retrognathic maxillae, increased mandibular planes angles, large gonial angles, and markedly negative apical base angle than milder cases with Binder's who were successfully treated orthodontically.

As most patients with Binder's syndrome requiring or requesting orthodontic treatment will be under the care of a plastic/maxillofacial surgeon, it would be sensible to have an inter-disciplinary approach to treatment planning throughout their care.

References

1. Munro IR Maxillonasal dysplasia (Binder's syndrome). *Plas Reconst Surg* 1979; **41**: 536–542.
2. Holmstrom H. Kahnberg K. Surgical approach in severe cases of maxillonasal dysplasia (Binder's syndrome). *Swed Dent J* 1988; **12**: 3–10.

3. Demas PN, Braun TW. Simultaneous reconstuction of maxillary and nasal deformity in a patient with Binder's syndrome (Maxillonasal dysplasia). *J Oral Maxillofac Surg*, 1992; **50**: 83–86.
4. Noyes FB. Case report. *Angle Orthod* 1939; **9**: 160–5
5. Binder KH. Dysostosis maxillo-nasalis, ein archinencephaler Missbildungskomplex. *Deutsche Zahnarztuche Zeitschift* 1962; **17**: 438–44.
6. McCollum AGH, Wolford LM. Binder Syndrome: Literature review and long-term follow-up on two cases.*Adult Orthod Orthognath Surg* 1998; **13**: 45–58
7. Eliasson LA, Holmstrom H. Midface prognathism in maxillonasal dysplasia (Binder's syndrome) determined by a theoretical midface ratio. *Int J Adult Orthodon Orthognath Surg*. 1994; **9**(4): 291–301.
8. Olow-Norderam M, Radberg CT. Maxillonasal dysplasia (Binder syndrome) and associated malformations of the cervical spine. *Acta Radiologica Diagnosis* 1984; **25**: 353–360.
9. Horswell BB, Holmes AD, Barnett JS, Levant BA. Maxillonasal dysplasia (Binder's Syndrome): A critical review and case study. *J Oral Maxillofac Surg* 1987; **45**: 114–122.
10. Olow-Nordenram M,Thilander B. The craniofacial morphology in persons with maxillonasal dysplasia (Binder syndrome). *Am J Orthod Dentofac Orthop* 1989; **95**: 148–58.
11. Ferguson JW, Thompson RPJ. Maxillonasal dysostosis (Binder syndrome) a review of the literature and case reports. *Eur J Orthod* 1985; **7**: 145–148.
12. Olow-Nordenram M, Valentin J. An etiologic study of maxillonasal dysplasia-Binder's syndrome. *Scand J Dent Res* 1987; **96**: 69–74.
13. Olow-Norderam M. Maxillonasial dysplasia (Binder's syndrome). A study of Craniofacial morphology, associated malformations and familial relationships. *Swed. Dent. J. Suppl.* 1987; 471–38
14. Gorlin R, Pindborg JJ, Cohen M Jr. Maxillonasal dysplasia (Binder syndrome). Syndromes of the head and neck. 1976 2nd Edition New York: McGraw-Hill
15. Olow-Nordenram M, Thilander B. The craniofacial morphology in individuals with maxillonasal dysplasia (Binder's syndrome). A longitudinal cephalometric study of orthodontically untreated children. *Eur J Orthod* 1987; **9**: 224–236.