

Masticatory dysfunction in persons with Down's syndrome.

Part 2: management

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SUMMARY The functional and anatomical characteristics of Down's syndrome have direct repercussions on oral health; orofacial dysfunction results and feeding and swallowing are impaired. These problems have been described in an earlier article. Different techniques are proposed for the prevention of the development of orofacial dysfunction in Down's syndrome. In particular, early myofunctional therapy coupled with appliance wear has been shown to be successful over the long term when multidisciplinary management is possible. Functional or conventional orthodontic treatment may be successful for older children when performed

concurrently with the use of appropriate behaviour management techniques. More recently, techniques for the compensation of masticatory dysfunction in adults have been proposed, although further research is necessary to confirm their efficacy. The aim of this second article was to review techniques for the prevention, treatment and compensation of orofacial dysfunction in persons with Down's syndrome from birth to adulthood.

KEYWORDS: Down's syndrome, mastication, deglutition, adults, children

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Introduction

A detailed description of the aetiology of orofacial dysfunction is given in the first article of this series, but briefly, the characteristics of Down's syndrome include dysfunction on account of poor neuromotor control (1), dental anomalies, orofacial dysmorphology and intercurrent illness (2). Anatomically, the facial mid-third is underdeveloped but the mandible follows normal development (3). The oral and facial musculature, in particular the tongue and lips are hypotonic. The tongue gives the impression of being abnormally large on account of muscle weakness and an anterior, low position in the mouth ('relative macroglossia') (4, 5). The temporomandibular joints are reported to be lax (2, 6). Masticatory dysfunction results and feeding problems are reported for persons with Down's syndrome at all stages of life. These problems may have important medical and social repercussions for persons with Down's syndrome and should be prevented wherever possible.

Prevention of orofacial dysfunction in infants and children with Down's syndrome has been the subject of much research over the last 25 years (6–11). Different techniques of myofunctional stimulation and appliance therapy have been proposed, and results have generally been positive provided the treatment is commenced at a very young age and if multidisciplinary collaboration is systematic (between speech therapists, physiotherapists, ENT specialists and dentists, etc.) (6, 12–23). For older children, conventional orthodontic treatment may also be successful when multidisciplinary cooperation allows. In adults with Down's syndrome, solutions to reduce masticatory dysfunction by compensating for marked malocclusion have only recently been proposed (24–26). These techniques have shown initially positive results but further research is necessary to evaluate their true impact on feeding. Despite the interest shown in the literature regarding these problems, few dentists are trained to diagnose, predict, prevent and treat masticatory dysfunction in this population.

The aim of this second article was to review the techniques for the prevention, treatment and compensation of masticatory problems within the population with Down's syndrome at different stages of development to encourage dentists to get involved in this important aspect of patient management.

Prevention and therapy for babies with Down's syndrome

Although neonates are rarely seen in the dental surgery, successful prevention of orofacial dysfunction starts from birth. Breast feeding may be initially difficult for babies with Down's syndrome but can be successful if appropriate encouragement is given (27, 28). For bottle-fed babies, bottles are sometimes adapted with an enlarged hole in the teat to decrease the force needed to feed. This practice should be avoided if possible as the orofacial musculature needs to be strengthened by suckling and there is a risk of choking (7). Techniques for improving suckling include gentle facial massage before feeding, ensuring that the tongue is retracted before giving the teat using the caregiver's fingers to ensure an oral seal around the teat, placing the index finger under the chin to aid swallowing and applying intermittent traction to the bottle (7). Nasogastric feeding in the neonatal period is sometimes necessary, but great care must be taken to ensure that the suckle-swallow reflex is stimulated during feeding or it may be lost. Appropriate stimulation is to give the baby a pacifier/dummy or a finger to suck on while nasogastric feeding is taking place, or to gently stroke the face and tongue (7). If effective suckle-swallow function is established this will help to reduce later stage problems by increasing orofacial muscle strength, stimulating maxillary skeletal growth and ensuring coordination of the respiratory swallowing sequence.

Prevention and therapy for infants and children with Down's syndrome

From approximately 6 months, feeding techniques aimed at developing normal oral function can be taught to families (7, 27, 29–31). Encouragement may need to be given upon the introduction of solid food (32). A short, shallow, rounded spoon is recommended with only a small amount of food at each time. The spoon should be placed on top of the tongue with a slight downward pressure and the food should not be scraped off onto the gums (7). Sticky food or paste (such as

peanut butter) can be given and the child is encouraged to remove it all from the spoon. Chewy solids should initially be placed in the molar region of the mouth to encourage chewing action and finger foods are useful. Food in strips can be guided between the teeth from one side of the mouth to the other to encourage lateral chewing. On introducing fluids from a cup, a spout should not be used and care should be taken that the tongue is retracted on drinking, rather than wedged under the edge of the beaker (7, 31) (Figs 1a, b and c). For older children (age around 2–3 years), drinking through a straw should be encouraged (7, 31). Parents should also be shown how to clean the child's nostrils regularly until the child can blow his or her own nose to encourage nasal respiration.

A mature chewing–swallowing sequence should ideally have replaced the primary suckle-swallow reflex by the time the second primary molars reach occlusion. For many children with Down's syndrome, this will not be the case and oromotor therapy is indicated. Unfortunately, many of the different methods of therapy lack sufficient research to be evidence-based, although research interest has increased over the last 20 years. The aims of all the different techniques are similar and involve encouraging development of the oral structures and mature masticatory function by increasing muscle tonicity, stabilizing the mandible at rest, freeing mandibular movement and providing occlusal contact to stimulate the periodontal sensorimotor system (6, 8–11). Collaboration between dental professionals, physiotherapists and speech therapists is essential as appliance wear is combined with orofacial stimulation. In cases of marked mouth-breathing, ENT colleagues may also be solicited to advise on the need for tonsillectomy and/or adenectomy (33).

Early intervention oromotor therapy involves oral stimulation and pre- and post-speech therapy (6, 22, 32). Pre-speech therapy exercises are useful for infants and games involving the introduction of different objects, textures and temperatures into the mouth should be encouraged (7). Such techniques are also useful in gradually diminishing an anterior gag reflex. For some children, early stimulatory appliance therapy may be useful, for example using the Castillo-Morales plate (6, 12–16, 34). These techniques were introduced approximately 25 years ago (4, 34, 35) and consist of physiotherapy of the oral structures (22, 36) and appliance therapy to stimulate the lips and tongue. The palatal appliance has a small suction ring on the

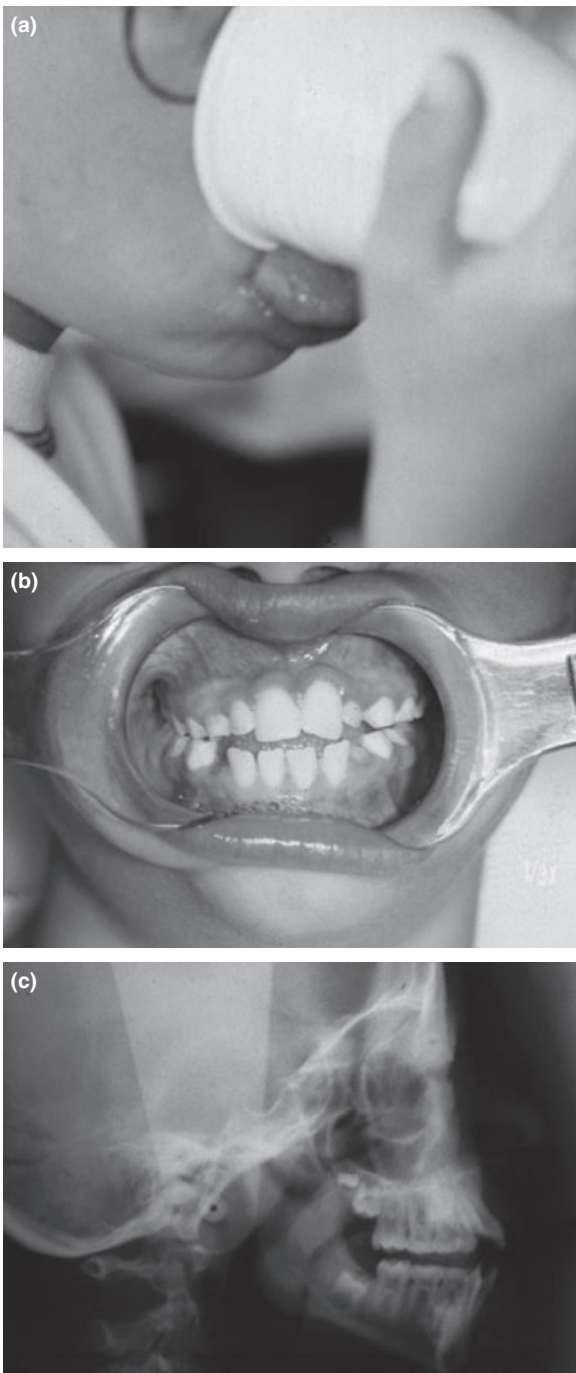


Fig. 1. Tongue position on drinking should be corrected. The consequences of uncorrected tongue protrusion are an anterior open bite and tipping of the incisors despite underdevelopment of the maxilla.

palate that encourages the child to raise his or her tongue and to perform oral movements. The plate may incorporate irregularities or a small lip bumper with beads on it, to encourage movements of the lip (Fig. 2).

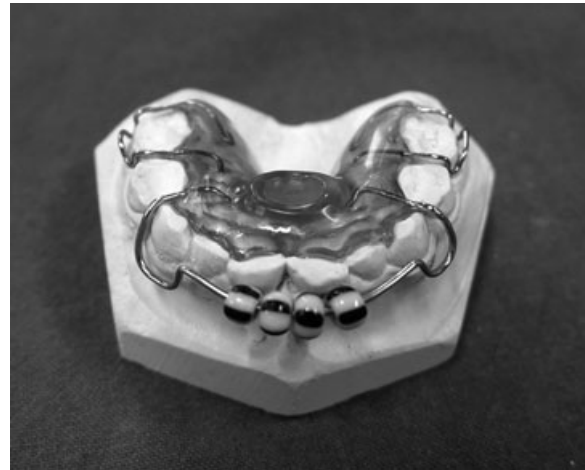


Fig. 2. Example of a stimulating palatal appliance – note suction cup and lip bumper.

The plate is worn for two or three periods of 15–60 min during the day – if left longer, it is no longer perceived as a foreign body and does not stimulate movement. The appliances are adapted regularly to accommodate growth and to provide stimulation as appropriate to the tongue and lips (12).

The results of this type of intervention have so far been very promising (6, 12–14, 18–23). The technique has been reported to help establish a rest position of the tongue behind the incisors, improve the position and strength of the lips, reduce tongue protrusion, encourage nasal breathing, reduce mandibular protrusion and correct the open mouth habit (8, 12, 37). The technique has also been reported to be helpful in eliminating drooling and improving swallowing and mastication (6, 8, 9, 12, 13). The results are variable depending on the child's cooperation but the best results have followed very early intervention (between the age of 3 months and 4 years) (6, 12–23). This may be explained by the concept of a 'critical period' for the introduction of certain stimuli after which a particular pattern of behaviour becomes increasingly difficult to learn (38).

Oromotor therapy may also be combined with functional orthodontic techniques. Simple intervention therapy includes palatal expansion with removable appliances or planned extraction of deciduous teeth to allow the permanent teeth optimum space on eruption. Elimination of occlusal interferences by grinding (usually the canine cusps) (39) or addition of composite to occlusal surfaces may free mandibular movement and help to restore periodontal stimulation (40–43) (Fig. 3).



Fig. 3. Composite overlays may help to establish a stable, retruded mandibular position and free the occlusion, particularly, for children who are unable to accept removable appliances.

Myofunctional therapy may be coupled with appliances designed to help to reposition the tongue, improve tonicity and encourage nasal respiration, for example, a tongue ramp or lingual envelope that is worn at night and guides the tongue to a resting position behind the upper incisors (44, 45). Such therapy may also be useful in the long term to help prevent recession of orthodontic treatment (46).

Certain children with Down's syndrome are able to accept conventional fixed orthodontic treatment without difficulty, but the practitioner should bear in mind that there will be increased need for retention on account of the underlying neuromuscular aetiology of the malocclusion. Concomitant orofacial stimulation or myofunctional therapy should be used, in particular, to improve lingual posture and mobility. In very rare cases, where the patient is able and willing to fully cooperate and give informed consent, it may be possible to combine orthognathic surgery with conventional orthodontic treatment, although it should be borne in mind that early preventive therapy is always preferable.

Prevention and therapy for adults with Down's syndrome

For adults who have not benefited from early intervention, marked oral dysfunction and severe malocclusion are common. The ultimate aims of treatment will be the same as for younger patients but techniques aimed at compensating for dysfunction will usually be preferred (24–26). For certain adult patients, the provision of adapted bite-raising interocclusal appliances to compensate for malocclusion may be helpful (25, 26, 47, 48) and has been reported to be successful for small

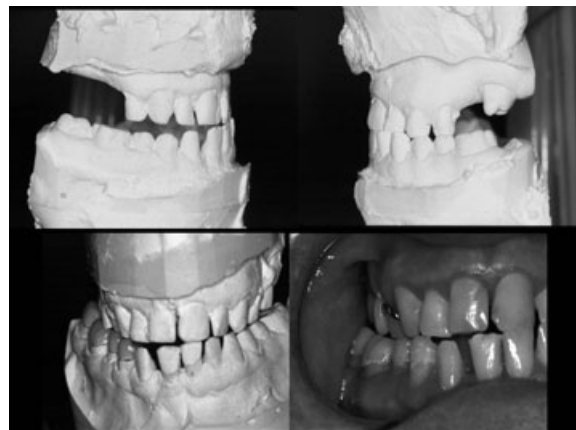


Fig. 4. This adult with Down's syndrome showed chronic luxation of the left temporomandibular joint. Fixed composite overlays were used to re-establish occlusal stability.



Fig. 5. Mandibular protrusion and tooth wear of the lingual surfaces of the lower incisors corrected using a removable bite-raising appliance. Following several months, the appliance shows wear facets that imply that the molars are now actively in occlusion.

groups of patients (24–26). Bite-raising appliances may be removable or fixed in the form of composite onlays (Figs 4, 5). The increase in vertical dimension seems well tolerated as it allows a degree of postural comfort absent from a marked malocclusion (24–26, 49). It seems that such appliances provide almost immediate improvement in comfort, aesthetics and in the reduction of dyskinetic movement and bruxism. It is likely, however, that to obtain lasting improvement in masticatory efficiency, oromotor stimulation therapy is also required to strengthen the oral musculature (26). A minority of patients may be amenable to adult orthodontic treatment, although usually the degree of discrepancy between the arches is too great for conventional treatment. In addition, unless muscular

function is corrected at the same time as the occlusion is established, orthodontic treatment is likely to require permanent retention.

In addition, correction of occlusal instability may be achieved by appropriate use of fixed and removable prostheses. This treatment has the advantage of replacing missing teeth and correcting occlusal discrepancies simultaneously (24, 25). Prosthetic rehabilitation with complete dentures is particularly successful as a stable occlusion is easier to achieve when replacing the entire dentition, especially in the presence of marked skeletal mismatch. Such treatment seems generally well accepted by patients with Down's syndrome as oral comfort is improved.

Patient management issues

It cannot be sufficiently emphasized that the functional problems encountered in children with Down's syndrome do not resolve with age, and on the contrary are likely to get worse if not corrected. The need for early familiarization of the child with Down's syndrome with the dental environment is highlighted to be able to intervene as soon as the primary dentition is in place and/or as the permanent dentition becomes established (50). Therapy is easier to establish at a young age, when parents and child are often already involved in specific educational programmes including speech therapy. Most children are sufficiently cooperative to accept simple appliance therapy if time is taken and appropriate behaviour management techniques are used. Conscious sedation techniques such as inhalation of nitrous oxide in oxygen may be necessary to perform treatment (to take impressions, for example) but as the child is seen regularly a relationship of confidence between child and dentist is established. Use of general anaesthesia and intravenous sedation for orthodontic intervention in the population with intellectual disability have been reported but remain controversial (50–53). Intervention needs to be long term and regular but treatment should take into account the individual's life project and social circumstances. Therapy should not be allowed to take a disproportionate place in the child or parent's life (54), and the parents or child should not be allowed to 'fail' a treatment attempt – for example if the child is unable to cope with therapy then treatment should be postponed and regular reviews maintained. Issues of cooperation and consent are more complex for the adult population, and treatment objectives may

have to be limited to meet the desires of the patient. Practitioners need to be careful to differentiate the wishes of the patient from those of their parents or caregivers. They also need to be aware that persons with Down's syndrome tend not to express pain or discomfort (55). In all cases and at all ages, an evaluation should ensure that the texture of food provided is adapted to the masticatory ability of the individual. Parents and caregivers should be made aware of the difficulties experienced by their ward and given guidance on the appropriate presentation of food.

Conclusion

The prevention of problems of orofacial function is of paramount importance. It is essential that dentists involved in the management of these patients be aware of the impact of functional problems on daily life, and that they learn to diagnose, predict, prevent and treat problems as they arise. The neuromotor deficit associated with Down's syndrome leads to the problems of orofacial function described. The treatment must also therefore be based in this domain for it to be successful long term. Although certain preventive and therapeutic measures proposed have a reasonable level of evidence, further research is required to confirm the effectiveness of the different types of intervention. Such research would also aid our understanding of oral rehabilitation for all patients with masticatory dysfunction of whatever origin. Perhaps in this way the additional disability of masticatory deficiency can be avoided for future generations of persons with Down's syndrome.

References

1. Frith U, Frith CD. Specific motor disabilities in Down syndrome. *J Child Psychol Psychiatry*. 1974;15:293–301.
2. Hennequin M, Faulks D, Veyrune JL, Bourdiol P. Significance of oral health in persons with Down syndrome: a literature review. *Dev Med Child Neurol*. 1999;41:275–283.
3. Fischer-Brandies H. Cephalometric comparison between children with and without Down's syndrome. *Eur J Orthod*. 1988;10:255–263.
4. Hoyer H, Limbrock GJ. Orofacial regulation therapy in children with Down syndrome using the methods and appliances of Castillo-Morales. *J Dent Child*. 1990;57:442–444.
5. Dodd B, Leahy J. Down's syndrome and tongue size. *Med J Aust*. 1984;140:748.

6. Carlstedt K, Dahllöf G, Nilsson B, Modeer T. Effect of palatal plate therapy in children with Down syndrome. A one-year study. *Acta Odontol Scand.* 1996;54:122–125.
7. Arvedson J. Management of swallowing problems. In: Arvedson J, Brodsky L, ed. *Pediatric swallowing and feeding: assessment and management.* San Diego, California: Singular Publishing group Inc.; 1993:327–388.
8. Glatz-Noll E, Berg R. Oral dysfunction in children with Down's syndrome: an evaluation of treatment effects by means of videoregistration. *Eur J Orthod.* 1991;13:446–451.
9. Limbrock G, FischerBrandies H, Avallé C. Castillo-Morales orofacial therapy: treatment of 67 children with Down syndrome. *Dev Med Child Neurol.* 1991;33:296–303.
10. Connolly B, Morgan S, Russell F, Fulliton W. A longitudinal study of children with Down syndrome who experienced early intervention programming. *Phys Ther.* 1993;73:170–181.
11. Daglio S, Schwitzer R, Wüthrich J. Orthodontic changes in oral dyskinesia and malocclusion under the influence of myofunctional therapy. *Int J Orofacial Myology.* 1993;19:15–24.
12. Carlstedt K, Henningsson G, McAllister A, Dahllöf G. Long-term effects of palatal plate therapy on oral motor function in children with Down syndrome evaluated by video registration. *Acta Odontol Scand.* 2001;59:63–68.
13. Carlstedt K, Henningsson G, Dahllöf G. A four-year longitudinal study of palatal plate therapy in children with Down syndrome: effects on oral motor function, articulation and communication preferences. *Acta Odontol Scand.* 2003;61:39–46.
14. Carlstedt K, Henningsson G, Dahllöf G. A longitudinal study of palatal plate therapy in children with Down syndrome. Effects on oral motor function. *J Disabil Oral Health.* 2007;8:13–19.
15. Hohoff A, Ehmer U. Effects of the Castillo-Morales stimulating plate on speech development of children with Down syndrome. *J Orofac Orthop.* 1997;58:330–339.
16. Hohoff A, Ehmer U. Short-term and long-term results after early treatment with the Castillo-Morales stimulating plate. *J Orofac Orthop.* 1999;60:2–12.
17. Daikoku H, Amano A, Fukui N, Akiyama S, Morisaki I. Clinical evaluation of orofacial regulation therapy for Down syndrome children using Castillo-Morales palatal plate. *Pediatr Dent J.* 2000;10:133–137.
18. Schuster G, Giese R. Retrospective clinical investigation of the impact of early treatment of children with Down's syndrome according to Castillo-Morales. *J Orofac Orthop.* 2001;62:255–263.
19. Backman B, Grever-Sjolander AC, Holm AK, Johansson I. Children with Down syndrome: oral development and morphology after use of palatal plates between 6 months and 18 months of age. *Int J Paediatr Dent.* 2003;13:327–335.
20. Backman B, Grever-Sjolander AC, Bengtsson K, Persson J, Johansson I. Children with Down syndrome: oral development and morphology after use of palatal plates between 6 and 48 months of age. *Int J Paediatr Dent.* 2007;17:19–28.
21. Zavaglia V, Nori A, Mansour NM. Long term effects of the palatal plate therapy for the orofacial regulation in children with Down syndrome. *J Clin Pediatr Dent.* 2003;28:89–93.
22. Korbmacher H, Limbrock J, Kahl-Nieke B. Orofacial development in children with Down's syndrome 12 years after early intervention with a stimulating plate. *J Orofac Orthop.* 2004;65:60–75.
23. Korbmacher H, Limbrock J, Kahl-Nieke B. Long-term evaluation of orofacial function in children with Down syndrome after treatment with a stimulating plate according to Castillo-Morales. *J Clin Pediatr Dent.* 2006;30:325–328.
24. Faulks D, Veyrune JL, Hennequin M. Consequences of oral rehabilitation on dyskinesia in adults with Down's syndrome. A clinical report. *J Oral Rehabil.* 2002;29:209–218.
25. Mazille MN, Veyrune JL, Hennequin M. Compenser les grandes dysmorphies: le concept de la stabilisation de la mandibule. *Real Clin.* 2005;16:63–74.
26. Mazille MN, Woda A, Nicolas E, Peyron MA, Hennequin M. Effect of occlusal appliance wear on chewing in persons with Down syndrome. *Physiol Behav.* 2008;93:919–929.
27. Aumonier ME, Cunningham CC. Breast feeding in infants with Down syndrome. *Child Care Health Dev.* 1983;9:247–255.
28. Pisacane A, Toscano E, Pirri I, Continisio P, Andria G, Zoli B *et al.* Down syndrome and breastfeeding. *Acta Paediatr.* 2003;92:1479–1481.
29. Cullen S, Cronk C, Pueschel S, Schnell R, Reed R. Social development and feeding milestones of young Down syndrome children. *Am J Ment Defic.* 1981;85:410–415.
30. Sleight D, Niman C. Intervention strategies for children with down syndrome. In: Rines J, ed. *Gross motor and oral motor development in children with down syndrome. Birth to three years.* St Louis, MO: St Louis Association for Retarded Citizens Developmental Services Department, 1984:47–49.
31. Kelso RA, Price S. Activities during the pre-toddler and toddler period. In: Burns Y, Gunn P, eds. *Down syndrome: moving through life.* London: Chapman and Hall, 1993;38–45;65–66.
32. Hopman E, Csizmadia C, Bastiani W, Engels Q, De Graaf E, Le Cessie S *et al.* Eating habits of young children with Down syndrome in The Netherlands: adequate nutrition intakes but delayed introduction of solid food. *J Am Diet Assoc.* 1998;98:790–794.
33. Venail F, Gardiner Q, Mondain M. ENT and speech disorders in children with Down's syndrome: an overview of pathophysiology, clinical features, treatments, and current management. *Clin Pediatr.* 2004;43:783–791.
34. Castillo-Morales R, Crotti E, Avallé C, Limbrock G. Orofaciale Regulation beim Down-Syndrom durch gaumenplatte. *Sozial pädiatrie.* 1982;4:10–17.
35. Limbrock GJ, Hoyer H, Scheying H. Drooling, chewing and swallowing dysfunctions in children with cerebral palsy: treatment according to Castillo-Morales. *J Dent Child.* 1990;57:445–451.
36. Limbrock GJ, Castillo-Morales R, Hoyer H, Stöver B, Onufer CN. The Castillo-Morales approach to orofacial pathology in Down syndrome. *Int J Orofacial Myology.* 1993;19:30–37.
37. Dahan JS, Lelong O. Effects of bite raising and occlusal awareness on tongue thrust in untreated children. *Am J Orthod Dentofacial Orthop.* 2003;124:165–172.
38. Light J. Sensory/motor therapy for the treatment of oral dyskinesia. A new approach to the treatment of oromyofunc-

- tional disorders with the use of tactile cueing handheld exercisers. *Int J Orofacial Myology*. 1995;21:23–28.
39. Martin E. Rehabilitation neuro-occlusale et meulages selectifs: resultats à un an. *Orthod Fr*. 2000;71:57–60.
 40. Simoes WA. Selective grinding and Planas' direct tracks as a source of prevention. *J Pedod*. 1981;5:298–314.
 41. Canalda K, Planas P. Interception in malocclusion [French]. *Orthod Fr*. 1983;54:313–327.
 42. Planas P. Notre testament en rehabilitation neuro-occlusale. *Orthod Fr*. 1991;62:695–705.
 43. Planas P. Equilibre et rehabilitation neuro-occlusale. *Orthod Fr*. 1992;63:435–441.
 44. Strazielle C, Mahler P, Alard R. La rééducation linguale: une nouvelle approche. *Inf Dent*. 1997;7:401–407.
 45. Bonnet B. Un appareil de reposturation: l'enveloppe linguale nocturne (ELN). *Rev Orthop Dentofaciale*. 1992;26:329–347.
 46. Mason RM. Orthodontic perspectives on orofacial myofunctional therapy. *Int J Orofacial Myology*. 1988;14:49–55.
 47. Ramfjord SP, Ash MM. Reflections on the Michigan occlusal splint. *J Oral Rehabil*. 1994;21:491–500.
 48. Ash MM Jr, Ramfjord SP. Reflections on the Michigan splint and other interocclusal devices. *J Mich Dent Assoc*. 1998;80:32–35, 41–46.
 49. Ormianer Z, Gross M. A 2-year follow-up of mandibular posture following an increase in occlusal vertical dimension beyond the clinical rest position with fixed restorations. *J Oral Rehabil*. 1998;25:877–883.
 50. Becker A, Shapira J. Orthodontics for the handicapped child. *Eur J Orthod*. 1996;18:55–67.
 51. Hobson RS, Nunn JH, Cozma I. Orthodontic management of orofacial problems in young people with impairments: review of the literature and case reports. *Int J Paediatr Dent*. 2005;15:355–363.
 52. Becker A, Shapira J, Chaushu S. Orthodontic treatment for disabled children – a survey of patient and appliance management. *J Orthod*. 2001;28:39–44.
 53. Chaushu S, Gozal D, Becker A. Intravenous sedation: an adjunct to enable orthodontics treatment for children with disabilities. *Eur J Orthod*. 2002;24:81–89.
 54. Sloper P, Cunningham C, Arnljotsdottir M. Parental reactions to early intervention with their Down's syndrome infants. *Child Care Health Dev*. 1983;9:357–376.
 55. Hennequin M, Morin C, Feine JS. Pain expression and stimulus localisation in individuals with Down's syndrome. *Lancet*. 2000;2:1882–1887.

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