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Mandibular resorption due to systemic sclerosis

Case report of surgical correction of a secondary open bite deformity

P. E. Haers, H. F. Sailer
Department of Maxillofacial Surgery,
University Hospital, Zurich, Switzerland

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Abstract. Systemic sclerosis (SSc) is a connective-tissue disorder of unknown origin causing a multisystem disease. While erosions of the distal phalanges are commonly described, resorption of the mandible has been considered an unusual finding. However, systematic radiographic screening of different groups of patients suffering from SSc revealed a resorption incidence of 20–33% of the examined mandibles. Women especially seem to be affected, and the male/female ratio is 1/7. Bilateral condylysis due to SSc has been described in seven cases, or 13.7% of the reported cases. To the best of our knowledge, this is the fourth report of surgical correction of secondary dysgnathia due to systemic sclerosis and the first with a 2-year follow-up period.

Key words: systemic sclerosis; mandibular resorption; condylysis; apertognathia; orthognathic surgery.

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Systemic sclerosis (SSc) is a generalized disorder of small arteries, microvessels, and diffuse connective tissue, characterized by fibrosis and vascular obliteration in the skin, gastrointestinal tract, lungs, heart, and kidneys. Hidebound skin is the clinical hallmark, and organ compromise the prognostic keystone[17]. Discontent has arisen concerning the use of the term "progressive systemic sclerosis" (PSS) largely because the disease is not always progressive, and calling it so places a serious emotional burden on the patient, and because SS without P may be confused with Sjögren's syndrome[17]. The disease is relatively uncommon with an average annual incidence of 6–12 new patients per million population and a prevalence of 130 per million[21]. It is most commonly diagnosed between the third and fifth decades. SSc rarely begins during infancy or after age 60 years, and women are affected about seven times as often as men[38]. SSc is an uncommon disease of uncertain origin and has been classified both as a "collagen-vascular" disease and an "autoimmune" disease. Genetic, environmental, and autoimmune factors are all supposed to be involved[7,13,15,41,43].

The basic pathogenesis is believed to be vascular injury of small arteries[17]. It often starts with the development of Raynaud's phenomenon. Characteristic of SSc is the tight firm skin which may be present several years before visceral involvement becomes apparent. Other clinical features are swelling of joints and ulceration of digits, muscular atrophy, flexion contracture of the hands, and rheumatic pain. Calcific deposits may develop in the subcutaneous and periarticular tissue, sometimes perforating the skin. The fibrosis involves not only the skin but also various internal organs, mostly the gastrointestinal tract, lungs, heart, and kidney. Resorption of the terminal phalanges of the hand and the distal portions of the radius and ulna are the most frequent radiologic findings in SSc patients[18]. Resorption of the ribs and distal clavicular resorption have been described as well[2]. Rheumatoid factors are present in 25% and antinuclear antibodies in 50% of the patients. Anti-Scl70 (in 15–40% of patients) and anticentromere (in<5–50%) are specific for SSc.

Diagnosis is straightforward. A previously well person develops Raynaud's phenomenon, nonpitting edema, and hidebound skin eventually covering virtually the entire body except the back and buttocks[17]. This sclerodermatous involvement proximal to the metacarpal-phalangeal or metatarsal-phalangeal (MCP) joints is the major criterion for diagnosis of diffuse SSc (dSSc) and provides the definitive diagnostic criterion of SSc in over 90% of the patients[20]. When distal to the MCP joints only, it is called limited SSc (lSSc) and is not diagnostic for SSc but can as well be suggestive of a spectrum of



Fig. 1. Hands of SSc patient are deformed, showing atrophic skin, cyanotic, ulcerated fingertips, and degenerated nails.



Fig. 2. A) Preoperative frontal view characterized by mask-like appearance with sclerotic skin, pinched nose, and severe atrophy of lips and perioral soft tissues. Muscular atrophy is clearly visible. B) High angle profile without well-defined chin, and retrusion of mandible.

scleroderma-associated conditions. The histology of nailfold capillary dilation, endothelial destruction, and capillary dropout (only in the diffuse form) is characteristic.

Classification in scleroderma includes a clear distinction between generalized (dSSc) and localized disease (lSSc), largely because internal or visceral disease does not occur in the localized form[17]. This last category is proposed to include and to replace the CREST variant of SSc (*C*alcinosis, *R*aynaud's phenomenon, *E*sophageal hypomotility, *S*clerodactyly, and *T*elangiectasia). Males exposed to chemicals, plastics, vibrating machines, or mining may develop lSSc which is often serologically negative[3]. No single drug or combination of drugs has proved satisfactory in the treatment of SSc in suitably controlled prospective trials[4]. The most promising results have probably been obtained with D-penicillamine (Mercaptyl®)[12]. Cyclosporin A has been used in a few patients with positive results[4]. Among Caucasians, the prognosis is worse in men than in women, but prognosis tends to be more severe in blacks, especially in black females. The disease progresses chronically, interrupted by partial spontaneous remissions with improvement of the skin. Patients with mainly skin involvement have a more gradual and favorable course than those with visceral disease[1].

The oral and maxillofacial manifestations of SSc are the mask-like appearance, muscular atrophy, thin atrophied lips, limitation of mouth opening, secondary microstomia, and rigidity of tongue and lips[6,9,19,23,38]. Widening of the periodontal ligament space is found radiographically in most patients[19,44]. Resorption of the mandible has been described as well and will be discussed more extensively.

Clinical report

Patient history

A 28-year-old Caucasian woman complained of altered facial appearance with worsening open bite and mandibular retrusion, which she had noticed during the previous 3 years. Photographs taken when she was 19 years old already depicted slight retrusion of the mandible in comparison with pictures from 2 years earlier at age 17, when she first suffered from Raynaud's phenomenon. Until the age of 21, this situation worsened with arthritic degeneration of the fingertips and periods of gastrointestinal involvement and generalized rheumatic pain, necessitating several hospitalizations. At the age of 22, SSc was diagnosed and the patient underwent cortisone therapy. Five years later, resorption of the distal phalanges and of the left mandibular condyle was noticed, while the sclerotic generation of the skin involved the lower arms and the shoulders.

Clinical examination

The hands were deformed, showing atrophic skin, cyanotic, ulcerated fingertips, and degenerated nails (Fig. 1). The facial appearance was characterized by atrophy of the skin and the facial muscles, affecting mostly the lips and the mentalis muscle (Fig. 2A and B). Retrusion of the mandible was evident, and the lower facial third was long. The upper incisors were exposed, the lips were extremely thin, and there was an interlabial gap of 15 mm. Mobility of the temporomandibular joints was reduced with maximal mouth opening of 34 mm. On palpation, the masticatory muscles were weak. There was a severe Class II occlusion with an anterior open bite of 8 mm and an occlusal contact limited to the right second molars (Fig. 3). Model analysis and abrasion of the cuspids indicated that a Class II occlusion with occlusal contact in the molar and bicuspid area and articulatory contact with the canines had existed in the past.

Radiologic examination

The orthopantomogram revealed bilateral condylysis and resorption of the coronoid processes, the posterior borders of the ascending rami, and the gonial areas. The periodontal ligament space in the premolar-molar area was widened (Fig. 4). The cephalo-

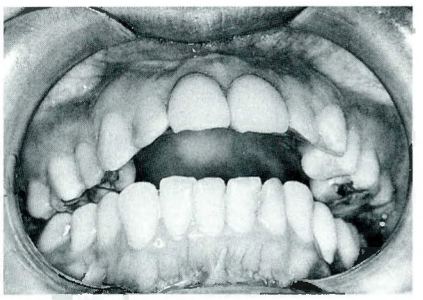


Fig. 3. Bilateral condylysis has caused frontal open bite with minimal occlusal contact in area of right second molar.

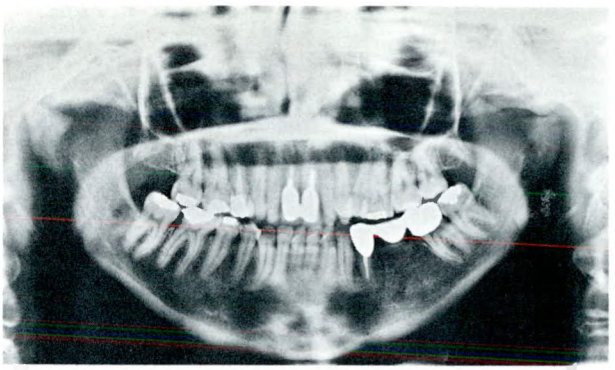


Fig. 4. Orthopantomogram revealing mandibular resorption: bilateral total resorption of condyles, and partial resorption of coronoid processes and posterior borders of ascending rami and gonial areas. Note widening of periodontal space ligament in bicuspid-molar area.

gram showed a high angle profile without well-defined chin configuration and a short mandible.

Surgical treatment

As the patient requested surgical correction and since her general condition had remained relatively stable during an observation period of 3 years, she was operated 14 years after the onset of disease. A maxillary intrusion of 7 mm combined with 10 mm advancement of the mandible and an advancement genioplasty of 7 mm were performed[24]. Further advancement of the chin was impossible because of lack of elasticity of the soft tissues. Fixation of the segments was performed with bank-bone interpositioning and wire osteosynthesis in the upper jaw, compression screws at the level of the sagittal split, wire osteosynthesis in the mental area and intermaxillary fixation with additional mandibular fixation using paranasal wires. Healing was normal and the mandibular and intermaxillary fixation was removed after 6 weeks.

Results

Although an acceptable result was obtained (Fig. 5), the occlusion was not stable. Three months postoperatively, there was a relapse of 2 mm, and 6 months postoperatively, an overjet of 6 mm and an overbite of 1 mm were seen (Fig. 6A). Two years postoperatively, an Angle Class II occlusion with nonocclusion of both canines was observed (Fig. 6B). The maximal mouth opening was 42 mm. The skeletal relapse was accompanied by further sclerosis of the soft tissues, especially in the mental area (Fig. 7). Cephalometric analysis from 3 years preoperatively until 2 years postoperatively showed gradual opening of the mandibular angle, suggesting ongoing resorption of the cranial part of both rami with subsequent shortening of the ascending ramus (Fig. 8A and B). Orthopantomographic tracings made during the 3-year preoperative and the 2-year postoperative period confirm this ongoing resorption pattern (Fig. 9). However, the result obtained was more functional compared to the preoperative situation, as occlusion was still present in the bicuspid and molar area.



Fig. 5. Profile 2 years postoperatively, showing improvement of nasolabial angle, shortening of lower facial third, and corrected chin contour.

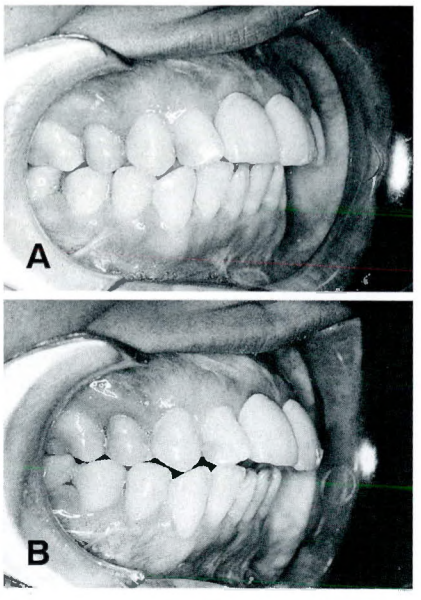


Fig. 6. A) Occlusal view 6 months and B) 2 years postoperatively, showing ongoing Class II/open bite relapse.

Discussion and literature review

Twenty-two publications reporting about 52 mandibles with different resorption patterns due to SSc were analyzed (Table 1). In 1949, atrophy of the ascending ramus following trauma was reported in a case where scleroderma was locally present[36]. Mandibular resorption due to SSc was first described by TAVERAS in 1959[35]. Several case reports mentioning unilateral partial or total resorption of the condyle as well as other mandibular resorption patterns have since been published (Table 1). In two cases, the zygomatic arch also was involved[6,9]. Fracture of a mandible weakened by resorption due to SSc has been reported twice[34,39]. Successful corticancellous grafting has been reported in a patient suffering from SSc and osteomyelitis of the mandible following nonunion of a fracture which the patient had sustained 11 years earlier[10]. Similar patterns of chronic mandibular resorption have been seen in a case of vinyl chloride acro-osteolysis. The patient developed Raynaud's phenomenon and all other features of SSc after a 6-year-long professional exposure to polyvinyl chloride. Radiographs of the mandible showed erosion of the posterior borders, complete loss of the mandibular angles, and erosion of the left condyle and of the right coronoid process[11]. Bilateral condylysis due to SSc has been described in seven cases or 13.7% of the reported cases of mandibular resorption[5,16,19,25,27,28,37].

Several authors have reported the incidence of mandibular resorption they found during systematic investigation of groups of SSc patients[2,19,34,42,44] (Table 2). They found a greater incidence of progressive mandibular resorption than individual case reports implied, because this phenomenon was detected in 20–33% of the patients.

Analysis of all cases of mandibular resorption reported reveals the follow-



Fig. 7. Frontal view A) 6 months and B) 2 years postoperatively, demonstrating progressing soft-tissue atrophy, especially in mental area.

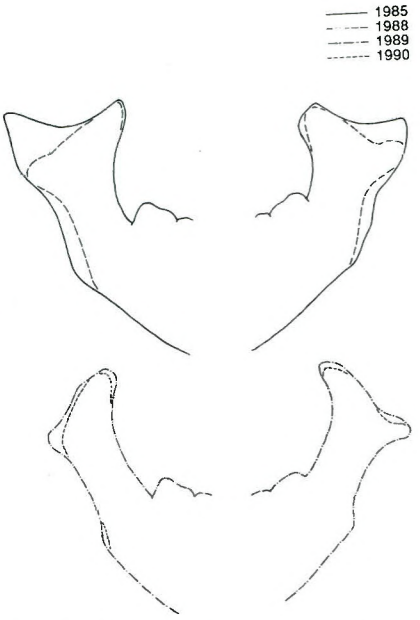


Fig. 9. Evolution of both ascending rami over 7-year period, as seen on orthopantomogram. Preoperatively, there was considerable resorption of cranial part of ascending rami which was continuing gradually, postoperatively.

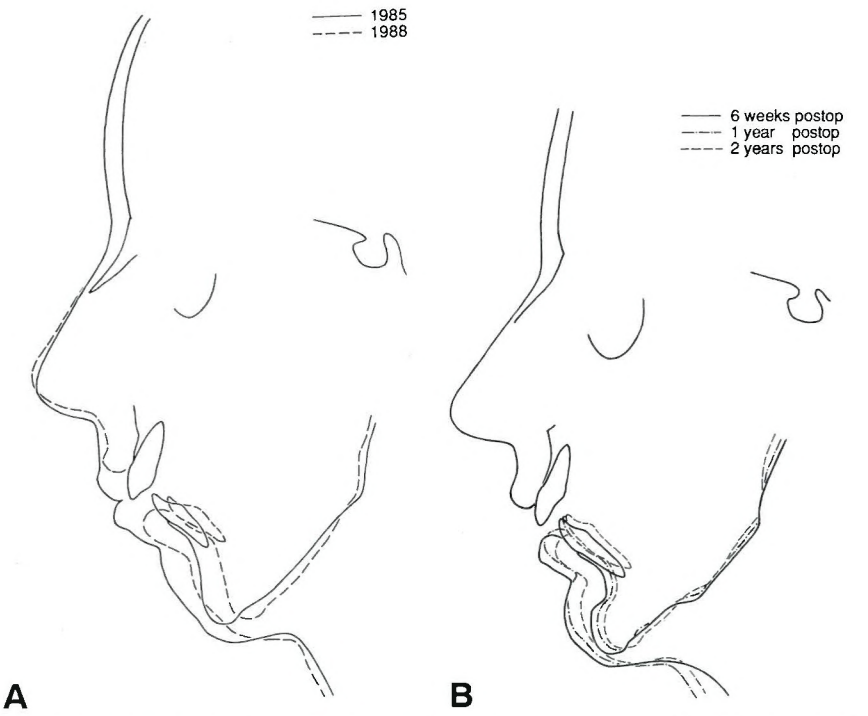


Fig. 8. Cephalometric tracings A) 3 years and immediately preoperatively and B) 8 weeks, 1 year, and 2 years postoperatively, illustrating ongoing opening of gonial angle due to further resorption of cranial part of ascending rami.

ing data. The mean age of these patients is 40 years with a minimum of 14 and a maximum of 62 years. The male/female ratio is 1/7. Most commonly, the resorption is detected between the fifth and the seventh years after SSc has been diagnosed. Among the reported cases of progressive mandibular resorption, the mandibular angle is involved most often (37.6%), followed by the condyle (20.8%), the coronoid process (20%), and the posterior border of the ascending ramus (14.4%). Resorption in other areas is rare.

The bone resorption is thought to be due to both pressure ischemia and vascular ischemia; the former is caused by the tight sclerotic facial skin and muscular dystrophy, the latter by obstruction of the small muscular blood supply. The loss of mandibular bone seems to be associated with muscular attachments[38]. As the blood supply of the condyle, the coronoid processes, and the gonial angles comes from small muscular arteries, it is suggested that the obstruction of these vessels causes the ischemic osteolytic lesions and the atrophy of the masseter and pterygoid muscles[28].

The correlation between the incidence of progressive mandibular resorption and the severity, duration, and multiorganic involvement of the disease is unclear. MARMARY et al.[19] could not establish a correlation between the incidence of progressive mandibular resorption and the incidence of widened periodontal ligament width, which is found in most patients with a long history of dSSc. In their series, patients with an extremely widened periodontal space did not present severe jaw erosion, while four patients with minor widening of the periodontal space suffered from gross mandibular resorp-

Table 1. List of cases reported with mandibular involvement in SSc (see references)

Author(s)	Age (years)	Sex	Years of SSc	Resorption pattern
TAYLOR[36] (1949)	19	M	5	Resorption of posterior border of ascending ramus
TAVERAS[35] (1959)				Right condylysis; erosion of medial portion of left condyle; bilateral resorption of mandibular angle
WEBER et al.[39] (1970)	28	F	7	
SEIFERT et al.[34] (1965)	62	F	5	Erosion along superior portion of condyles and unilateral angular resorption
	49	M	3	Flattening of condylar heads and minimal bone loss at both angles
	39	F	13	Bilateral blunting condyles and severe bone loss at both angles
	61	F	11	Flattening of left condyle and bilateral moderate resorption of angle
	31	F	7	Bilateral absence of coronoid complex, only small twig of bone from condule to angle
RABEY[27] (1977)	14	M	5	*Bilateral condylysis* and antegonial resorption
WHITE et al.[42] (1977)				Unilateral resorption of angle
				Unilateral destruction of coronoid process
				Bilateral angular resorption
				Bilateral angular resorption
				Bilateral angular resorption
				Bilateral resorption of coronoid process and angle
				Bilateral resorption of coronoid process and angle
LANIGAN et al.[16] (1979)	26	F	6	*Bilateral condylar resorption;* wide glenoid fossa; erosion of anterior border of ascending ramus
BASSETT et al.[2] (1981)				Bilateral angular resorption
				Bilateral angular resorption
				Bilateral angular resorption
				Bilateral angular resorption
				Bilateral resorption of angles and coronoid process
				Bilateral resorption of angles and coronoid process
				Bilateral coronoid resorption
MARMARY et al.[19] (1981)	27		4	*Bilateral total condylysis* and total resorption of collum
	28		9	*Bilateral total condylysis* and total resorption of collum
	39		3	Unilateral resorption of coronoid
	53		11	Bilateral resorption of coronoid and posterior border
OSIAL et al. [25] (1981)	26	F	5	*Bilateral resorption of condyle and coronoid process*
WEINER & WOLF[40] (1981)	49	F	9	Bilateral erosion of angle and ramus
HOPPER & GILES[9] (1982)	59	F	33	Bilateral absence of mandibular angle and most of ascending ramus
NAYLOR[23] (1982)	59	F	5	Bilateral antegonial resorption
RAMON et al.[28] (1987)	22	F	1	*Bilateral condylysis*
WARDROP & HEGGIE[38] (1987)	22	F	4	Bilateral resorption of coronoid
POGREL[26] (1988)	59	F		Unilateral osteolysis of angle and coronoid
SCUTTELARI et al.[33] (1988)	61	F	11	Bilateral resorption of posterior border of ascending ramus
	32	F	3	Unilateral resorption of angle and ascending ramus
	55	F	6	Bilateral resorption of angle and ascending ramus
WOOD & LEE[44] (1988)				Nine patients with mandibular resorption patterns
RUPRECHT et al.[31] (1990)	55	F	7	Bilateral absence of coronoid and unilateral erosion of angle and condyle
CARTIER & BEZIAT[6] (1990)	54	F	7	Bilateral resorption of posterior border of ascending ramus
THALLER & KAWAMOTO[37] (1990)	32	F	7	*Bilateral loss of condyle, coronoid, and almost entire superior ramus*
	25	F		*Bilateral condylysis*
RUBIN & SANFILIPPO[30] (1992)	44	F		Bilateral resorption of angle

Reports on cases of bilateral condylysis causing open-bite deforming are indicated by italics.

Table 2. Systematic studies of mandibular resorption patterns in SSc patients

Author(s)	No. of patients	Age (years)	Male/female ratio	Years of SSc	Widened periodontal space	Mandibular resorption
SEIFERT et al.[34] (1975)	16	17–65		1–15; 7	3/11	5/16=31%
WHITE et al.[42] (1977)	35	24–79	5/30		13/35	7/35=20%
MARMARY et al.[19] (1981)	21	27–63		3–49; 13	21/21	7/21=33%
BASSETT et al.[3] (1981)	55	26–70	7/48	2–35; 11		7/35=20%
WOOD & LEE[44] (1988)	31	51.9	0/31	9.5		9/31=29%

Years of SSc: minimum–maximum; average.

tion. They also found that no correlation existed between mandibular resorption and age, duration of disease, medication, or clinical and laboratory findings. This contradicts the statement of Wood & Lee that mandibular erosions occurred in patients who had more severe systemic sclerosis with greater restriction of mouth opening and more widespread organ system involvement[44].

Although mandibular resorption and erosion can be seen in other conditions such as malignoma, metastasis, and infection, the resorption patterns in these cases are not localized around muscular insertions, as is the case in SSc, and they have different radiologic characteristics. However, a special condition which may resemble the resorption pattern seen in SSc is the so-called (Gorham's) massive osteolysis[8].

Condylar resorption and condylysis[17] have been described in association with several other pathologic conditions. Partial resorption and subcortical lucencies are known as part of the radiologic features of temporomandibular arthrosis and have been reported in association with rheumatoid arthritis. Partial dissolution also can occur followed by ankylosis[35]. Avascular necrosis of the condyle, too, can lead to erosions and resorption of the condyle[29,32]. Severe resorption of the condyle has also been reported to occur occasionally following orthognathic procedures[14,22].

In three cases, malocclusion due to condylysis induced by SSc was surgically corrected, one case being a mixed collagen vascular disease[16,37]. None of these reports described the postoperative course. Although it is mandatory to ascertain that condylar resorption has stopped in patients for whom orthognathic surgery is contemplated, resorption continued in the case presented, worsening the open bite and the lip incompetence. By performing an intrusion of the maxilla, eliminating the open bite, and thus reducing the vertical facial dimensions, it was hoped that the mechanical constriction caused by the rigid sclerotic musculocutaneous facial envelope would be reduced. Although lip competence and occlusion were improved, resorption of the condyles did not stop.

Nevertheless, the authors believe that surgery should not be refused in such cases, because a successful result will not only improve function but will also lighten the psychological and emotional burden of a deteriorating condition.

Postsurgical relapse suffered by one patient does not mean that all patients will relapse after surgery; however, the possibility of such negative postsurgical changes, whether secondary to the primary disease process or to the osteotomy itself, must play a prominent role in the planning of the treatment, and informed consent is mandatay.

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Address:
Dr P. E. Haers
Department of Maxillofacial Surgery
University Hospital Zürich
CH 8091 Zürich
Switzerland

This book is dedicated to the authors' spouses,
Martha, Peter, and Marilyn,
and to their children,
Samantha, Amanda, Blake, and MacKenzie;
Caroline and Amanda;
and Tiffany and Gabriel.

William Hoffman, M.D.
Associate Professor, Department of Surgery, Division of Plastic and Reconstructive Surgery, University of California at San Francisco School of Medicine, San Francisco, California
Comprehensive Surgical and Orthodontic Management of Hemifacial Microsomia

Leonard B. Kaban, D.M.D., M.D.
Walter C. Guralnick Professor and Chair, Department of Oral and Maxillofacial Surgery, Harvard School of Dental Medicine; Professor and Chair, Department of Oral and Maxillofacial Surgery, Massachusetts General Hospital, Boston, Massachusetts
Comprehensive Surgical and Orthodontic Management of Hemifacial Microsomia

Sven Kreiborg, D.D.S., Ph.D., D.Odont.
Professor, School of Dentistry, Faculty of Health Sciences, University of Copenhagen, Copenhagen, Denmark
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Clinical Nurse Specialist, Craniofacial Center, Children's Hospital Oakland, Oakland, California
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