

Correction of Facial Deformity in Sturge–Weber Syndrome

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Background: Although previous studies have reported soft-tissue management in surgical treatment of Sturge–Weber syndrome (SWS), there are few reports describing facial bone surgery in this patient group. The purpose of this study is to examine the validity of our multidisciplinary algorithm for correcting facial deformities associated with SWS. To the best of our knowledge, this is the first study on orthognathic surgery for SWS patients.

Methods: A retrospective chart review included 2 SWS patients who completed the surgical treatment algorithm. Radiographic and clinical data were recorded, and a treatment algorithm was derived.

Results: According to the Roach classification, the first patient was classified as type I presenting with both facial and leptomenigeal vascular anomalies without glaucoma and the second patient as type II presenting only with a hemifacial capillary malformation. Considering positive findings in seizure history and intracranial vascular anomalies in the first case, the anesthetic management was modified to omit hypotensive anesthesia because of the potential risk of intracranial pressure elevation. Primarily, both patients underwent 2-jaw orthognathic surgery and facial bone contouring including genioplasty, zygomatic reduction, buccal fat pad removal, and masseter reduction without major complications. In the second step, the volume and distribution of facial soft tissues were altered by surgical resection and reposition. Both patients were satisfied with the surgical result.

Conclusions: Our multidisciplinary algorithm can systematically detect potential risk factors. Correction of the asymmetric face by successive bone and soft-tissue surgery enables the patients to reduce their psychosocial burden and increase their quality of life. (*Plast Reconstr Surg Glob Open* 2016;4:e843; doi: 10.1097/GOX.0000000000000797; Published online 15 August 2016.)

Sturge–Weber syndrome (SWS) is a rare sporadic neurocutaneous disorder with a reported incidence of 1/50,000.¹ The disease is characterized by facial capillary and vascular malformations involving facial bones and soft tissue, eyes, and intracranial structures.² Since its proposal as a fourth phakomatosis entity in 1937, there has been a lot of debate on the

pathophysiology of SWS.^{3–5} A new hypothesis attributes localized primary venous dysplasia or acquired venous obstruction rather than malformation of the embryonic vascular plexus in the mesencephalic neural crest to the development of SWS.^{6,7} The consequent effect of focal venous hypertension leads to hypertrophy of the adjacent structures such as the calvarial bone, leptomenigeal and choroidal veins, and facial skin and bones. Prominent features such as capillary malformations, redundant skin, and hypertrophic facial bones result in regional disfigurement and dysfunction paired with dental malocclusion. This may adversely affect social development in terms of normal peer relationship, intelligence, or educational and professional success^{8,9} and may also translate into significant psychologic burden and social problems affecting the patient's quality of life.^{9,10}

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Several reports have described the surgical treatment for SWS and its associated problems, including ophthalmologic surgery for glaucoma and cataract caused by malformation of the anterior chamber or high episcleral venous pressure,^{11,12} orthodontic treatment for periodontal overgrowth and malocclusion,^{13–16} hemispherectomy for epilepsy caused by hemispheric pathology,^{17,18} and laser or surgical resection for capillary malformations and redundant soft tissue.^{10,19–22} The main anesthetic concern in SWS is airway management because of facial and airway hypertrophy, which can potentially hinder mask ventilation and laryngeal mask or endotracheal tube insertion. In addition, it is crucial to prevent a rise in intraocular and intracranial pressure to minimize the potential risk of seizure during anesthesia induction by appropriate doses of antiepileptics.

Considering the perioperative risks, surgical treatment for the facial deformity is challenging in SWS patients. Studies on facial bone surgery are limited.²¹ However, careful preoperative evaluation and a well-organized multidisciplinary approach can balance the potential risks and benefits of facial bone surgery and subsequently help decrease the patient's psychologic burden.

Our treatment strategy for addressing facial disharmony consists of 2 steps. The first step is to correct the underlying skeletal and dental abnormality using orthognathic surgery combined with facial bone contouring. Simultaneous contouring surgery can address the prominent structures including zygoma, chin, and the mandibular angle. To the best of our knowledge and from our literature review, this is the first study of orthognathic surgery for SWS patients. In the second step, the volume and position of facial soft tissues are altered by surgical resection and reposition. The purpose of this study was to present our multidisciplinary algorithm for correction of facial deformity associated with SWS with the outcome report of the first 2 patients.

MATERIALS AND METHODS

This retrospective study received approval of our institutional review board (number: 104-8604B). A retrospective chart review included SWS patients who completed the surgical treatment algorithm. The surgeries were conducted by the senior surgeon (L.J.L.) at the Craniofacial Center, Chang Gung Memorial Hospital, between October 1, 2011, and August 31, 2015. Exclusion criteria included the following: patients who had not completed the algorithm, a follow-up of less than 6 months, and incomplete or missing data. Two patients who met these criteria were included in the analysis. Patient demographics such as sex, Roach classification, age at the time of each surgery, type of surgery, operative time, intraoperative blood loss, amount of blood transfusion, and follow-up period were collected.

Treatment Algorithm

Preoperative Evaluation

After thorough history taking and physical examination, magnetic resonance imaging or contrast computed tomography (CT) angiography is conducted for

surgical candidates to detect possible intracranial lesions. If positive findings come up in central nervous system imaging or the patients have a seizure history, further evaluation of possible epilepsy requires referral to a neurologist or neurosurgeon. Ophthalmologic workup is necessary to detect glaucoma from possible choroidal venous hypertension. If patients have a history of epilepsy or glaucoma, the perioperative anesthetic management should be adjusted to minimize the potential risk for seizure. This includes the choice of appropriate anesthetic agents, anticonvulsants, and pharmacologic manipulation related to intraocular and intracranial pressure. A preoperative multidisciplinary team meeting should be held to assess the surgical feasibility considering the potential risks (Fig. 1).

Surgical Planning

Cone beam CT plays an important role in 3-dimensional (3D) simulation surgery, which can provide detailed skeletal morphology and measurement for 3D planning. The 3D simulation provides the precise position for the maxillary and mandibular segments.²³ Facial bone contouring for the zygoma, mandibular angle, and chin can also be outlined in the 3D plan for the facial asymmetry correction.

Surgical Technique—Orthognathic Surgery

Under general anesthesia with nasotracheal tube intubation and controlled hypotension, epinephrine-containing local anesthetic solution is injected for the mucosal incision and for regional blocking of the greater palatine and inferior alveolar nerves. In our department, single-splint 2-jaw surgery is the standard procedure, and both jaws are osteotomized and moved as 1 maxillomandibular complex to the intended position. A bilateral sagittal split osteotomy for the mandible and a LeFort I osteotomy for the maxilla are performed in the standard fashion. After osteotomy completion, maxillomandibular fixation is performed using the final occlusal splint. The maxillomandibular complex is mobilized with 3D translation and rotation to the new position as a single unit. The bony segments are fixed with 4 titanium plates for the maxilla, and 3 transbuccal bicortical screws on each side for the mandible. As indicated, simultaneous mandibular angle reduction is accomplished under direct vision before fixation. In most cases, genioplasty can balance the aesthetic E-line and the midline.²⁴ The horizontal osteotomy is performed 8mm below the mental foramen from a lower vestibule incision. After confirmation of the correct advancement and midline position of the distal segment, rigid fixation is performed with 2 titanium plates. Adjunct procedures such as zygomatic reduction can be performed in combination with buccal fat removal and masseter muscle reduction to control the soft tissue discrepancy on the hypertrophic side.²⁵

Surgical Technique—Soft-tissue Reduction with Reposition

In the second stage, it is important to assess the soft-tissue distribution in the sitting position to take gravity effects into account before the surgery. Under general

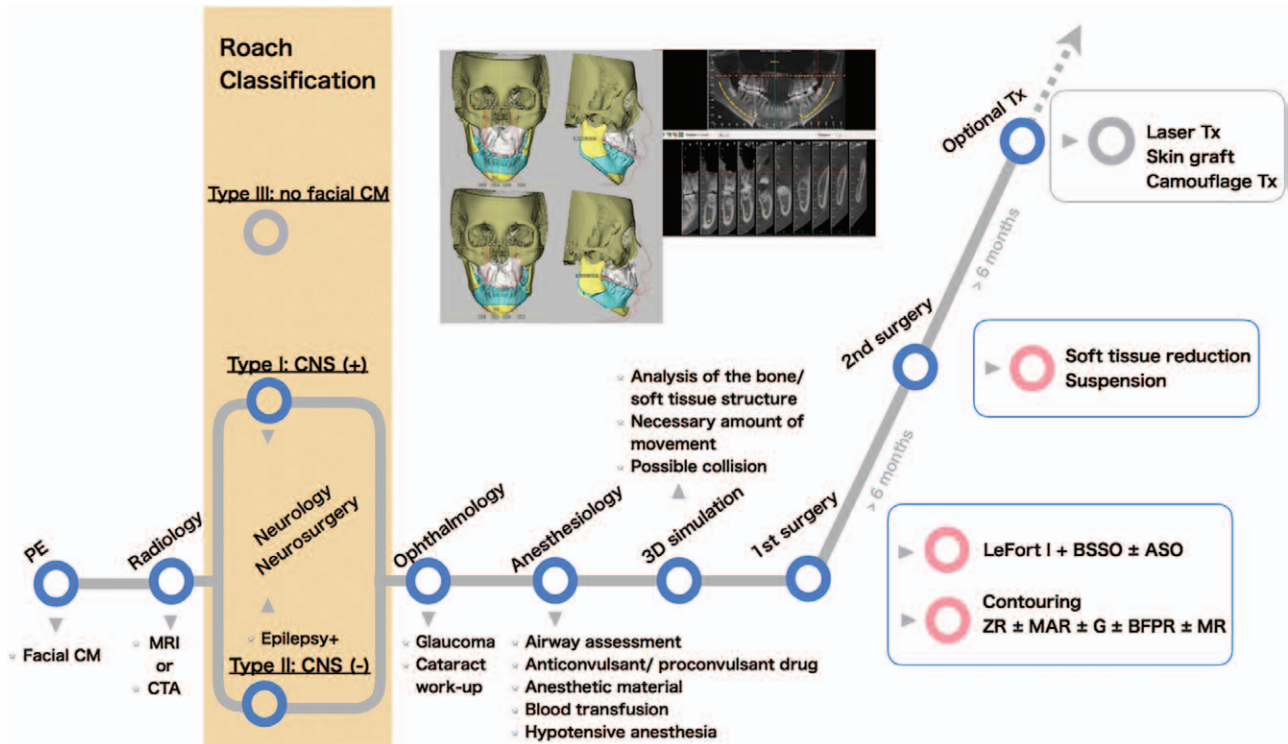


Fig. 1. Treatment algorithm for patients with Sturge–Weber syndrome. ASO indicates anterior segmental osteotomy; BFPR, buccal fat pad removal; BSSO, bilateral sagittal split osteotomy; CM, capillary malformation; CNS, central nervous system; CTA, computed tomography angiography; G, genioplasty; MAR, mandibular angle resection; MR, masseter reduction; MRI, magnetic resonance image; PE, physical examination; Tx, treatment; ZR, zygoma reduction; 3D, 3 dimension.

anesthesia, epinephrine-containing local anesthesia solution is injected before skin incision. Following the facial aesthetic units, a limited tissue dissection is made with enough thickness on the superficial layer to avoid surface irregularities. Simple excision of the redundant skin and subcutaneous tissue can balance facial harmony; however, sagging effects might be missed in a supine position. Anchoring sutures to underlying fixed structures such as periosteum or bone is mandatory to prevent postoperative drooping. Although intraoperative bleeding is limited and easily controlled with electrocautery, small drainage tubes may be helpful to reduce postoperative hematoma.

RESULTS

Patient demography including sex, Roach classification, age at the time of each surgery, type of surgery, operative time, intraoperative total blood loss, and follow-up period is described in Table 1. There were no major complications in the perioperative period of any of the surgeries.

Case 1

A 29-year-old woman had a congenital capillary malformation on the right side of her face. The contralateral side of the forehead was also involved in a skip lesion pattern (Fig. 2). Patient and family history revealed that she had

Table 1. Perioperative Data

Case	Gender	Roach Classification	Epilepsy	Age 1 (y)	Surgery 1	Operative Time (min)	Blood Loss (ml)	Blood Transfusion	Age 2 (y)	Surgery 2	Follow-up Period (mo)
1	F	I	Remission	31.0	LeFort I with ASO BSSO G Right ZR Both MAR Right BFPR	426	2,250	4 units RBC 1 unit WBC	32.6	Resection Suspension	49.4
2	F	II	No	15.8	LeFort I BSSO G Left ZR BFPR	195	1,740	4 units RBC	16.3	Resection Suspension	14.0

ASO, anterior segmental osteotomy; BSSO, bilateral sagittal split osteotomy; BFPR, buccal fat pad removal; CM, capillary malformation; CNS, central nervous system; F, female; G, genioplasty; MAR, mandibular angle resection; MR, masseter reduction; PE, physical examination; RBC, red blood cell; WBC, whole blood cell; ZR, zygoma reduction.



Fig. 2. A and B, A 29-year-old woman had a capillary malformation on the right side of her face. The contralateral side of forehead was also involved in a skip lesion pattern. C and D, After skeletal correction, the incisor show was adequately reduced. Total facial harmony was improved by soft-tissue reduction surgery.

2 seizure events at the age of 2 and 10 years. The seizure was not specified in detail; however, a course of antiepileptic drugs at the time of occurrence succeeded to induce remission and was withdrawn without any recurrence of seizures. There was no history of neurologic and psychiatric symptoms in the past 2 decades, even though CT angiography showed an extensive congenital intramedullary venous anomaly and

dystrophic calcifications flecking over the right frontal and left posterior subcortical regions. Diffuse, prominent, deep intramedullary veins drained into the choroid plexus and deep venous systems of the right atrium and the left paracavernous region (Fig. 3). Regarding facial bone structures, the imaging study showed hypertrophy of the zygoma and maxilla on the affected side with engorged veins (Fig. 3). Facial soft tissue was noticeably redundant on the affected cheek and upper lip area. She presented with class 2 malocclusion consisting of deep overbite because of regional osseous overgrowth, occlusal canting, and excessive gingival display even with the lip in the resting position (Fig. 4).

The patient was bronchoscopically intubated with a nasotracheal tube and placed under sevoflurane–fentanyl general anesthesia. Blood pressure was controlled by adjusting narcotics concentration and inhalation agents instead of using an adrenergic antagonist. Controlled hypotensive anesthesia was not performed because of the intracranial venous anomaly.

A combined approach with orthognathic and bone contouring surgery was performed, including tooth extractions (teeth nos. 18, 14, and 24), LeFort I and anterior segmental osteotomy of the maxilla, bilateral sagittal split osteotomy of the mandible, genioplasty, right zygoma reduction, and right buccal fat pad reduction. The intraoperative blood loss was 2,250 ml. There was no postoperative complication except for postoperative nausea and vomiting. Reduction of redundant cheek and upper lip soft tissue combined with upper lip suspension was performed 6 months later to balance facial harmony. The patient was satisfied with the surgical improvement.

Case 2

A 15-year-old girl had a capillary malformation stain affecting the left facial hemisphere from lower eyelid to upper lip (Fig. 5). She had several laser and sclerosing injection treatments for the facial venous lesions before presenting at our clinic. However, there was no history of neurophthalmologic or endocrine disorders. She presented with gradually enlarging left cheek soft tissue and overgrowth of the left maxilla, which caused anterior open

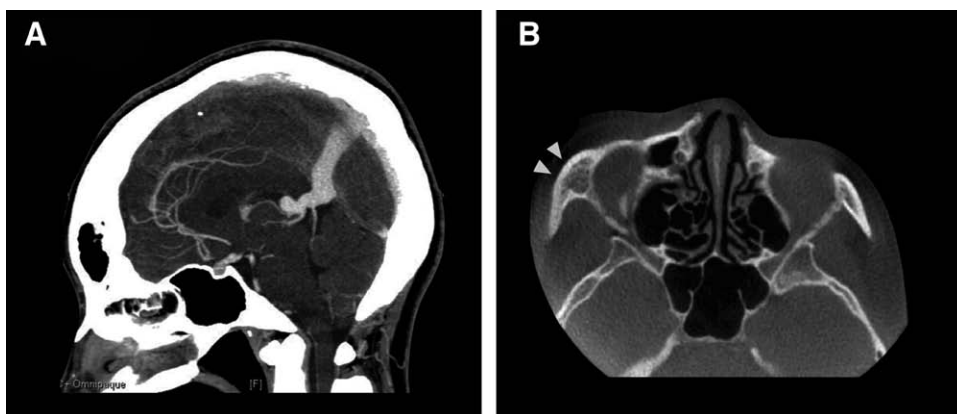


Fig. 3. Intracranial venous anomaly and calcification in CT. A, Sagittal section showed calcification in right frontal subcortical regions. Enlarged ventricular vein drained into sagittal sinus. B, Mild hypertrophy of zygoma and maxilla on the affected side with engorged veins.

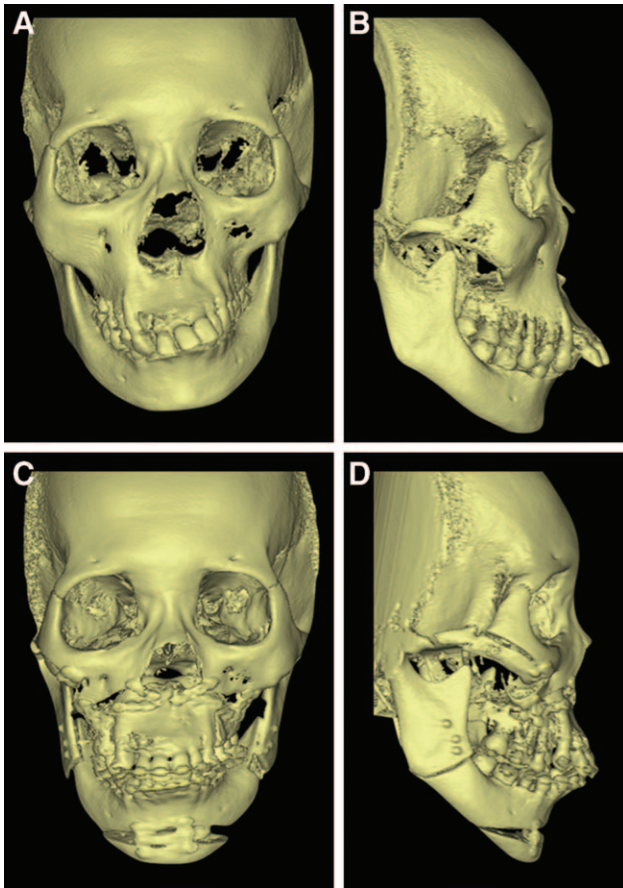


Fig. 4. A and B, Preoperative CT showed class II malocclusion with deep overbite, occlusal canting, and larger malar prominence on the right affected side because of maxillary hyperplasia. C and D, Malocclusion and occlusal canting were corrected by 2-jaw surgery with maxillary anterior segmental osteotomy, and the right zygoma prominence and mandibular angle were also balanced.

bite, occlusal canting, and malocclusion. She also had a history of prolonged bleeding after tooth extraction at 10 years of age. CT angiography showed mild hypoplasia of the right maxilla and soft tissue without any abnormal intracranial vessels. Although the left zygoma and maxilla were hypertrophic, both regions had a thinner cortex and heterogenous bone density compared with the unaffected side (Fig. 6).

The patient was placed under general anesthesia using fiberoptic nasotracheal intubation. Hypotensive anesthesia with labetalol was used to reduce intraoperative blood loss and increase the quality and visibility of the surgical field. The mean arterial pressure was maintained at 50 to 60 mm Hg.

She underwent orthognathic surgery consisting of LeFort I, bilateral sagittal split osteotomy, and genioplasty, combined with facial contouring surgery (left zygoma reduction and buccal fat pad reduction; Fig. 7). The intraoperative blood loss was 1,750 ml. There was no postoperative complication. Six months later, she underwent soft-tissue reduction for the redundant cheek tissue from a nasolabial incision and for the upper lip from a mucosal

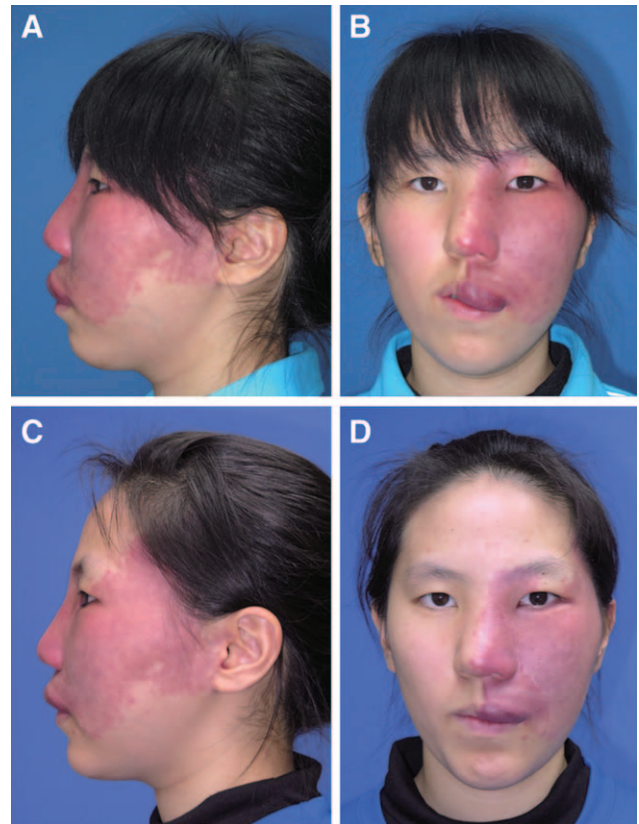


Fig. 5. A and B, A 15-year-old girl had a left capillary malformation ranging from the lower eyelid to the upper lip. C and D, Improvement of facial asymmetry was noted 6 months after procedures.

incision. The inferiorly positioned left nasal ala and upper lip were lifted to the piriform margin. She recovered well from both surgeries, with a substantial improvement of facial appearance, and was satisfied with the result.

DISCUSSION

Craniofacial problems of SWS patients can range from skin to brain disease of various degrees; because of the complicated pathophysiology, the surgical treatment of these patients requires a multidisciplinary team approach. The variegated hypertrophy of tissues can be explained with the increased venous and capillary transudate, leading to an increase in availability of extracellular nutrients and related growth factors and causing hypertrophy of soft and bony tissues. Gingival enlargement may also be present because of hypertrophy, antiepileptic medications such as phenytoin sodium, or a combination of both.²⁶ Facial bone overgrowth can cause severe facial disfigurement with facial and dental asymmetry, occlusal canting, increased dental show, and malocclusion. If aesthetic concern or malocclusion is beyond the limit of orthodontic treatment, orthognathic surgery is the only option for the correction of this deformity. Although many studies have reported surgical management related to periodontal or soft tissue, there are few reports describing facial bone surgery.^{10,16,26-28} The potential risks of massive bleeding, neurologic deterioration, intracranial

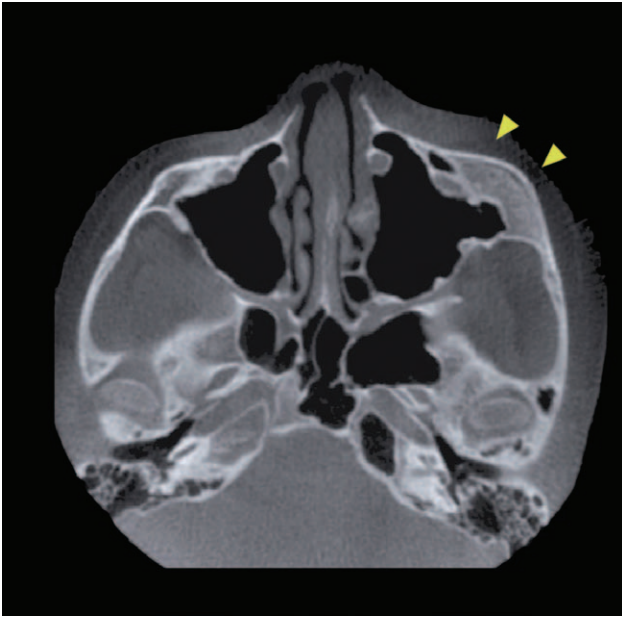


Fig. 6. The hypertrophic zygoma and maxilla on the left side were hypertrophic but with a thinner cortex and heterogenous bone density compared with the unaffected side.

al lesions, and anesthetic challenges including the higher complexity because of possible mental retardation may have led to a conservative indication policy for these patients.

Contrary to the limited studies on bone surgery in SWS, there are several studies about the surgical treatment for osseous venous malformations, which can also affect the size and vascularity of the facial bones.^{29–33} Two pathologic types for osseous venous malformations have been described: the central and the peripheral types. The central type arises from the cancellous vessels, whereas the peripheral type involves periosteal vessels and subsequently affects the bone. An en bloc resection can reduce massive intraoperative bleeding and recurrence of venous malformations especially for the central type. However, there may be an increased risk of morbidity associated with an extensive approach and complicated reconstructions.³⁰ Partial resection of osseous lesions can be an alternative with satisfactory results.^{33,34} In contrast, osseous hypertrophy in SWS was assumed to be derived mainly from local hypervascularity around either periosteum or spongiosa, or both.⁷ From this standpoint, combined orthognathic and contouring surgery in SWS could be regarded as a partial instead of complete resection of the lesion.

The critical anesthetic concern in SWS is airway management because facial and airway hypertrophy can hinder mask ventilation or endotracheal tube insertion. Our patients had no gross airway involvement; however, preoperative airway problems including obstructive sleep apnea should be evaluated.²⁷ The Roach classification is simple and convenient for categorizing clinical findings according to facial and central nervous system (CNS) involvement: type I consists of both facial and leptomeningeal vascular anomaly with or without glaucoma, type II

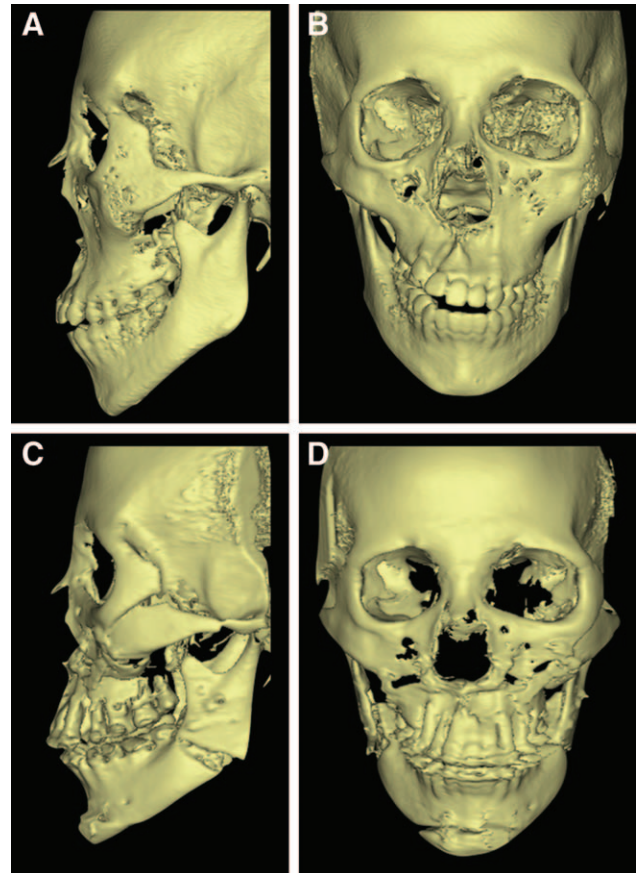


Fig. 7. Surface-rendered 3D images demonstrate the preoperative and postoperative skeletal structure. A and B, Preoperative CT showed mild hyperplasia of the left maxilla and mild hypoplasia of the right maxilla, which caused anterior open bite, occlusal canting, and malocclusion. C and D, Postoperatively the malocclusion and anterior open bite were corrected. Left zygoma was reduced to balance midface width.

presents with facial capillary malformations and no CNS involvement with or without glaucoma, and type III shows isolated leptomeningeal vascular anomaly without glaucoma.³⁵ Clinical CNS manifestations including seizures (23%–83%) and mental retardation (60%) are frequently observed in patients with type I or III anomaly.^{27,35} Although the first patient in this study had no CNS-related symptoms in the last 2 decades after the remission at the age of 10 years, it is necessary to inform the patient and family about possible perioperative and postoperative seizures and their potential deterioration.

If patients have a seizure history, the surgical indication must be judged through a thorough consideration of the potential risks and benefits including sudden death from severe seizure.³⁶ For anesthetic management, it is crucial to prevent a rise in intraocular and intracranial pressure in conjunction with minimal airway manipulation to prevent seizure attacks during anesthesia induction. Furthermore, the therapeutic level of antiepileptics and perioperative use of anesthetics with anticonvulsant property should be considered.²⁷ Disease progression has been linked to recurrent thrombosis and resulting venous

stasis. Low-dose aspirin use in patients with SWS has been reported to optimize neurodevelopmental outcome with minimal side effects because antiplatelet medications can reduce thrombosis and promote perfusion.³⁷ Perioperative indication for antiplatelet therapy should be carefully tailored weighing the possible merits for CNS perfusion and the risks such as bleeding or hematoma during epidural/spinal anesthesia.

Perioperative bleeding is one of the main concerns for any type of intervention. One reported mortality was attributed to uncontrollable bleeding from the diploic veins during burr-hole creation for a SWS patient.³⁸ Spontaneous intracerebral bleeding caused by sudden congestion of an intracranial vein by a venous thrombus has also been reported.³⁹ On the other hand, there are few reports describing massive bleeding in the surgical field. In our series, none of our patients received antiplatelet medications. However, we had a relatively large amount of blood loss because of continuous oozing from the affected bone. The intraoperative findings were compatible with the underlying etiology of SWS, which is not hyperactive blood inflow but congestion because of venous hypertension. Accordingly, the oozing from the affected bone was more severe compared with the nonaffected one. Considering the difficulty to control such oozing, it is better to shorten the total operative time to minimize blood loss. Three-dimensional simulation is a good option to reduce the operative time⁴⁰ because it allows detailed analysis of the complicated skeletal structure and an estimation of bony movement and possible collision in advance.²³ Furthermore, hypotensive anesthesia combined with epinephrine-containing local solutions is helpful to reduce perioperative blood loss.^{41–43} However, in case of intracranial involvement, hypotensive anesthesia with vasoactive agents might induce seizures because of elevation of intracranial pressure. Considering the adverse effects, we did not apply induced hypotensive anesthesia for the first case.

To the best of our knowledge, this is the first report of orthognathic surgery for correction of facial asymmetry in SWS. Aggressive treatment for SWS might have been avoided because of possible risks and complications, but addressing the stigmata and facilitating psychosocial support could be crucial to enhance the quality of life in patients with facial disfigurement.⁹ Our treatment protocol comprises 2 steps: the first step is focused on balancing the bony structures, and the second step on volume reduction and soft-tissue repositioning (Fig. 7). Ideal facial balance can be achieved combining maxillary and mandibular surgery instead of single-jaw surgery.⁴⁴ Additional bony deformities causing facial asymmetry can also be addressed at the same time using facial bone contouring surgery in combination with buccal fat removal and masseter reduction.²⁵

Although suction-assisted tissue removal was reported to be an effective and less invasive method for volume reduction even for patients with capillary malformation,²⁰ SWS involves subcutaneous and cutaneous tissue as well. Consequently, the soft-tissue redundancy of all layers has to be addressed to balance facial harmony. Several op-

tions such as laser therapy, skin grafting, and camouflage makeup have been reported for the color-patched appearance of the skin, but each method has its disadvantages and is unlikely to solve all problems on its own. More than 90% of capillary malformations may be refractory to laser therapy, but the combination of pulsed dye laser and topical sirolimus may improve the treatment success rate.⁴⁵ In contrast, surgical excision and coverage with skin grafts has been reported.^{46,47} Major disadvantages of skin grafts are donor site morbidity and possible secondary wound reactions such as hyperpigmentation and hypertrophic scarring.¹⁰ Consequently, cosmetic camouflage is still an effective option for patients with capillary malformation to treat the color disharmony, which can enhance patient's quality of life substantially.⁴⁸

Limitations to the present study exist. First, the limited case number prevents any conclusion about the procedures; however, this is the first report of orthognathic surgery for SWS and could, therefore, open up the possibility for SWS patients to choose corrective surgery for the asymmetric skeletal deformity. Second, the present cases are East-Asian patients, which could present regional difference of bleeding tendency, culture-based beauty concept, and motivation for treatment.⁴⁹ These issues warrant further investigation such as multicenter studies to collect a sufficient number of patients. However, we think our proposed treatment algorithm can systematically detect potential risk factors, and the results enable the patients to be more confident regarding their cosmetic appearance, reduce their psychosocial burden, and increase their quality of life.

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