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# Facial Duplication – The Unique Case of Antonio

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**Summary**

A case of facial duplication with its surgical correction in childhood and the consequences on facial growth is reported. It is a unique case in the duration of observation. The following structures were fully duplicated: the nose, the premaxilla, the cribriform plate, the crista galli. In addition there was an enormous facial cleft including lip, alveolus and palate. Additionally there were two rudimentary eye sockets, eyes, and two supplementary eyebrows. The monstrous hypertelorism with the facial duplication was corrected at the age of ten. The surgical procedure is described and the postoperative complications are discussed. Gross lack of growth of the middle third of the facial skeleton was observed. This was probably the consequence of the initial corrective surgery. Overgrowth of the mandible created a gorilla-like appearance by the end of the growth period. This was corrected in one operation by advancement of the middle third in three sections and repositioning of the mandible as a whole together with the mandibular anterior alveolar segment. Finally all parts of the lower half of the nose had to be enlarged, both soft tissues as well as the cartilaginous framework. A pharyngoplasty in addition to the correction of the intermaxillary abnormalities did much to improve the speech quality of the patient. A large secondary cranial defect was successfully reconstructed with the use of 14 halved ribs. In spite of the removal of four ribs from one side and three ribs from the other, there were no postoperative respiratory problems. Spontaneous rib regeneration was found where ribs had been removed one year earlier.

**Key-Words:** Facial duplication; Hypertelorism; Monstrous prognathism; Median facial cleft; Facial growth; Cranial defect reconstruction; Multiple resection of ribs; Duplication of premaxilla; Duplication of nose; Duplication of crista galli; Duplication of cribriform plate; Supernumerary eye sockets.

## Introduction

Antonio is unique in that he was born with two completely developed noses, an extremely wide median cleft of the face and palate, two complete premaxillae and a monstrous hypertelorism.

He is also unique in that we have learned more during his treatment than in any other case. We surgically corrected his craniofacial abnormality at the age of ten. By the age of eighteen, when his general growth had ceased, the development of the middle third of the face had lagged far behind that of the upper and lower thirds. This again caused a monstrous gorilla-like appearance in spite of an initially satisfactory result. This condition again made a gross movement of the skeletal structures of the middle and lower thirds of the face necessary. The growth disturbance was mainly attributed to the surgical intervention in childhood.

In yet another respect have we learned from Antonio: During the course of treatment we were confronted with numerous complications; their successful management led to unusual possibilities of treatment which were not immediately obvious to us.

Because Antonio is a unique case in various respects we believe it worthwhile to publish his history, especially as there is no case reported in the literature of duplication of the middle face which had been observed from the surgical correction in childhood until growth had ceased.

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Fig. 1



Fig. 2

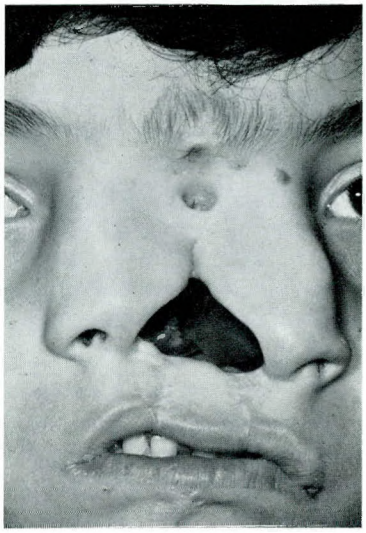


Fig. 3

Fig. 1 Antonio at the age of about 3 months. The tremendous median facial cleft and the 2 fully developed noses are the most striking parts of his facial duplication.

Fig. 2 Antonio at the age of ten. In early childhood the 2 median alae were approximated and a Abbé-flap was inserted into the upper lip and the soft palate and the dorsal half of the hard palate were closed elsewhere. The intercanthal distance of the hypertelorism measures 7.5 cm.

Fig. 3 Close up view of the medial facial section at the age of ten. The hypertrophic nasal conchae are visible between the joined medial alae. Between the two nasions is a groove following early removal of a growth of the size of a small cherry. According to reports it included two rudimentary eyes. Between the two eyebrows there is an additional third eyebrow.

**Previous History**

Antonio was born on 4th of May 1959. He presented an unusual face *(Fig. 1)*: There were two completely developed noses of normal size and shape with normal alae and nostrils. They were separated by tremendous median facial and palatal clefts. The eyes were set far apart and between the roots of the two noses was a pedunculated skin-covered growth the size of a small cherry. At the age of four months the pedunculated growth was removed. According to his mother's report the surgeon found two rudimentary eyes within the growth. At the age of eight months the medial alae of the two noses were approximated and at the age of 1 year the soft and hard palates were closed and later an Abbé-flap from the lower lip was used to close the wide gap in the upper lip.



Fig. 4 The state of the maxillae and the number of teeth at the age of ten: besides a large medial defect in the alveolus and anterior hard palate area there is a fully developed premaxilla included in each maxillary half.

According to his mother's report she had taken tablets during the pregnancy with Antonio but she does not remember their name. He was a forceps delivery.

There was no further surgery until he presented to us. He was referred to us for possible correction of his cranio-facial abnormalities by Prof. *Hugo Krayenbühl,* former chief of the Neurosurgery Clinic, University Hospital Zürich, to



Fig. 5 a



Fig. 5 b

Fig. 5 a + b Frontal tomograms of the facial skeleton at the age of ten: the two eye sockets are placed far laterally. This is also true of the maxillary sinuses. In a) a gross enlargement of the ethmoidal cell areas between the two eye sockets is striking and the same X-ray suggests the possible presence of two cristae galli. In b) there are two nasal cavities and two sphenoidal sinuses clearly visible. Each nasal cavity has a nasal septum as a medial wall. The two nasal septi form a roof to the tremendously enlarged oral cavity with the median cleft. The floor of the anterior cranial fossa reaches downwards almost to the level of the infraorbital rims.

whom he was sent because of his unilateral right sided impaired motor development.

### Clinical Findings

The boy was of normal general size for his age. The right arm and leg had grown less than the left extremities. There was a slight spastic hemiparesis on the right side. He had a limping gait. As a consequence of the hemiparesis there was a mild degree of thoracic kyphosis. All other general clinical findings were within normal limits.
Eye, ENT and speech findings are discussed in separate chapters.

Intellectual development was within normal limits. He had received his education partly from his mother at home, and partly in a community school. Emotionally, he was much disturbed by his facial deformity. Conscious of his malformation he was shy and avoided human contact. Speech quality was remarkably abnormal and almost unintelligible.

His appearance was dominated by the two almost complete noses with a deep groove between them and a defect above the upper lip which had an Abbé-flap inserted in the midline *(Fig. 2)*. On each side of that flap was a normal half of an upper lip including the philtrum and the shape of the vermilion. In the middle of the lower lip was a slightly hypertrophic scar obviously resulting from the Abbé-flap operation.

The tips of the two noses were 5.5 cm and the two nasions 4.5 cm apart. There was a normal lateral nostril and ala and a normal columella on each nose. The medial alae were somehow fused. Between them hypertrophic nasal conchae were visible *(Fig. 3)*. There was a groove between the two nasions, an obvious extra midline orbit,

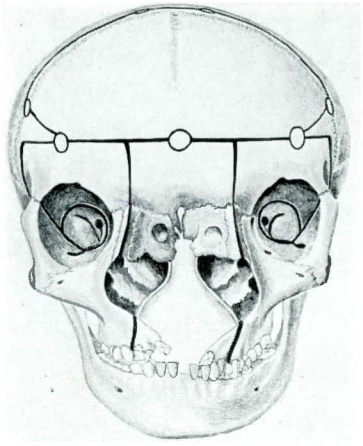


Fig. 6a

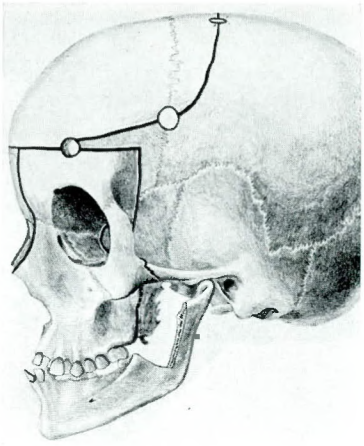


Fig. 6b

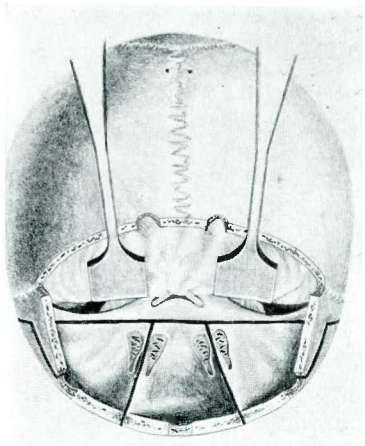


Fig. 6c

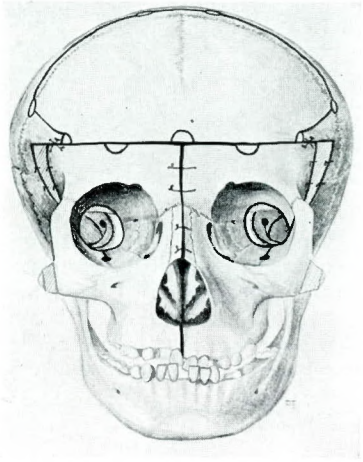


Fig. 6d

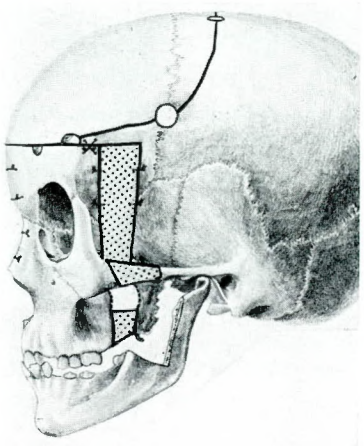


Fig. 6e

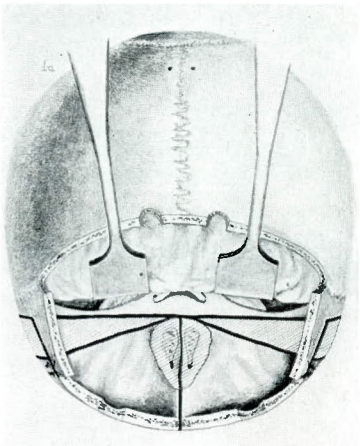


Fig. 6f

Fig. 6a–f Sketch of the planned operative procedure. a–c) The preoperative condition of the facial skeleton with the planned osteotomies. d–f) The facial skeleton after excision of a medial strip of the facial framework including a wedge shaped strip of the anterior cranial fossa and after medial rotation of the two facial halves. Areas with parallel oblique lines and dotted areas indicate bone grafts.

together with its eyebrow. There was an enormous hypertelorism with an intercanthal distance of 7.5 cm. There was conjugate movement of both eyes and the patient seemed to have normal vision (see ophthalmological findings).

Intraorally there was a wide median cleft in the middle of the upper alveolar process and the anterior part of the hard palate. The soft palate was closed, but scarred and short with limited movement. There was crowding of teeth in the anterior area of each maxillary alveolar process, presenting as a full premaxilla, as a part of the alveolar process of each side. On the left side there were four incisors, two completely developed central incisors of normal size and shape and two normal lateral incisors. On the right side one lateral incisor was missing, otherwise the teeth were the same as on the left side *(Fig. 4)*. In the anterior third of the hard palate and in the alveolus there was a large defect communicating with the oral cavity and with a large median cleft between the two nasal septi (see *Figs. 5b and 21a)*. There were some remaining deciduous teeth. Oral hygiene was very poor.

The lower jaw was of normal shape and size for the patients age. There was a normal number and size of teeth with some deciduous teeth still present. The intermaxillary relationship and cuspal interdigitation was almost normal laterally. There was, however, protrusion of the lower anterior teeth as easily seen in the preoperative cephalometric X-ray.

### X-ray Findings

The standard views and frontal tomograms show the enormous hypertelorism and the medial facial cleft *(Fig. 5)*. There are two sphenoidal sinuses. The floor of the anterior cranial fossa reaches far down between the two widely displaced orbital sockets. The radiologist suggested that there were possibly two cribriform plates. Carotid angiography of the left side showed abnormalities in the architecture and localisation of the cerebral vessels on that side. The anterior cerebral artery on the left side has at least three branches. The left lateral ventricle is enlarged compared with the normal.

### Plan of Treatment

Based on *Tessier's* (1967) transcranial procedure for the correction of monstrous hypertelorism the plan for the correction of this case was made *(Fig. 6 a–f)*. It was obvious that a medial strip of the facial skeleton had to be excised in order to permit the lateral facial halves including the eye sockets to be rotated medially and including also part of the adherent soft tissue structures all the way down from the forehead into the mouth, as well as everything medially from the top of the two nasal bony frameworks. It would also include a wedge shaped section of the anterior cranial fossa as far back and as close to the sphenoid limbus as possible. Both orbital coni would need to be osteotomized in their depth which means just in front of the supra-orbital fissure and optic foramen. On both sides a vertical osteotomy in the temporal fossa should be carried down through the lateral orbital wall and between the pterygoid process and the maxilla into the oral cavity. The zygomatic arch would be cut or the zygomatic-temporal suture. A large craniotomy would be used to obtain the necessary access to the anterior cranial fossa. Anteriorly protruding "fingers" of the frontal bones (*Tessier* 1967) would be left to permit direct bone wiring of the facial halves, after their medial rotation, to these, the only remaining stable pieces of the anterior craniofacial skeleton.

### The Operation itself

All doctors and nurses participating in the operation received a copy of the plan of operation laid down in detail covering 30 points. On the 4th July 1969 the operation was carried out step by step as planned preoperatively. General anaesthesia via an oro-tracheal tube was used. The prolonged anaesthesia was very satisfactorily maintained with a mixture of nitrous oxide, oxygen and methoxyflurane with controlled ventilation. Lumbar puncture and drainage was used to reduce intracranial pressure for the duration of surgery on the anterior cranial fossa. The frontal scalp was turned down from right to left temporal fossae. A large bilateral transfrontal craniotomy was performed. After exposure of the dura mater, the sagittal sinus was found to be situated to the right of the midline, starting above the base of the right nose. The left frontal lobe was found to be abnormally enlarged. The base of the anterior fossa was exposed extradurally. A small tear in the dura was sutured. In the anterior fossa two cristae galli were found, belonging to both noses. On each side of the crest the olfactory filaments penetrated the dura mater. There was no visible intracranial nervous connection to the two rudimentary eye sockets found between the two glabellas.

Through infraorbital incisions, an additional medial vertical scalp incision and an incision in the upper oral vestibular sulcus the periosteum and soft tissues were raised from the whole skeleton of the middle and upper thirds. A medial strip of bone was excised by osteotomies measuring 5 cm from one tip of the nasal framework to the other and 4.5 cm in height from the frontal bone *(Fig. 7)*; within the anterior cranial fossa the osteotomies passed posteriorly on top of the two cristae galli, ending 1 cm in front of the sellar tubercle. Here a horizontal osteotomy of 2.5 cm in length connected them. From the top of the nasal framework the osteotomy passed vertically down to the maxillary alveolar process and cut



Fig. 7 The two bony nasal frameworks before removal of the median skeletal strip of the face. The dark lines indicate the osteotomies. Laterally on both sides, the completely mobilized contents of the orbital coni are seen.

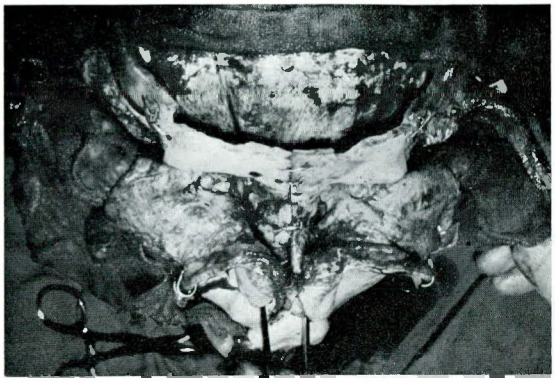


Fig. 8 After medial rotation of the facial halves a single bony nasal framework was created. Both rotated facial halves are already fixed to stable parts of the cranium.

off the premaxillary part on either side. The hard palate was incised in the midline.

The medial bloc of the facial framework could now be removed *(Fig. 9)*. It included almost the two complete premaxillae, the medial half of the nasal framework, the medial section of the forehead and below of it two rudimentary eye-sockets. It also included the upper and medial nasal conchae and both fully developed nasal septa. Furthermore it included a wedge-shaped piece of the anterior cranial fossa with each medial half of the two cristae galli and each medial half of the two cribriform plates. Damage to the brain, the optic nerves and the hypophysis was avoided. But the olfactory filaments were disrupted.

The periosteum of each orbit was raised close to the supraorbital fissure and optic foramen. Osteotomies were made in the depth of the orbital coni. After bilateral vertical osteotomies in the temporal fossae and lateral orbital walls and after separating the maxilla from the pterygoid process and an osteotomy through the zygomatic arch in the zygomatico-temporal fissure, both facial halves could be rotated medially. The rotation distance on each side measured 2.5 cm at the pupillary level. The facial halves were fixed together and to the "skull-fingers" in the new position by direct wires. Out of the two lateral halves of both skeletal nasal frameworks, one nasal skeleton of almost normal size, width and shape resulted *(Fig. 8)*. The interorbital distance measured 25 mm. The orbital coni, which were preoperatively at

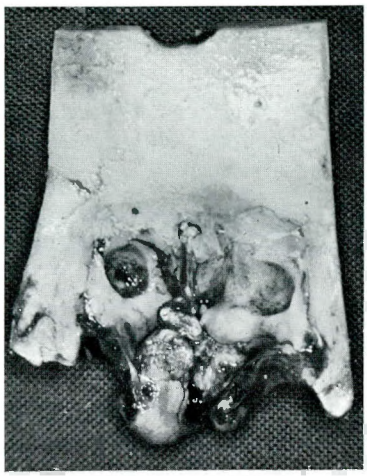


Fig. 9 Anterior aspect of the removed medial strip of the facial skeleton. It measured 5 cm between the tips of the two nasal frameworks and 4.5 cm in the height of the frontal bone. The additional two rudimentary eye sockets are easily identifiable.

the lateral edge of the skull, have been rotated to a normal position *(Figs. 10 a+b)*.

The bone defects in the anterior cranial fossa as well as in the temporal area and orbital coni were filled with bone grafts. To bridge the gap in the zygomatic arch a piece of a rib was wedged between the bone ends. The right iliac crest and three ribs from the right side were removed in order to provide the necessary amount of bone.

The exposed base of the skull including the grafted areas were covered with periosteum re-

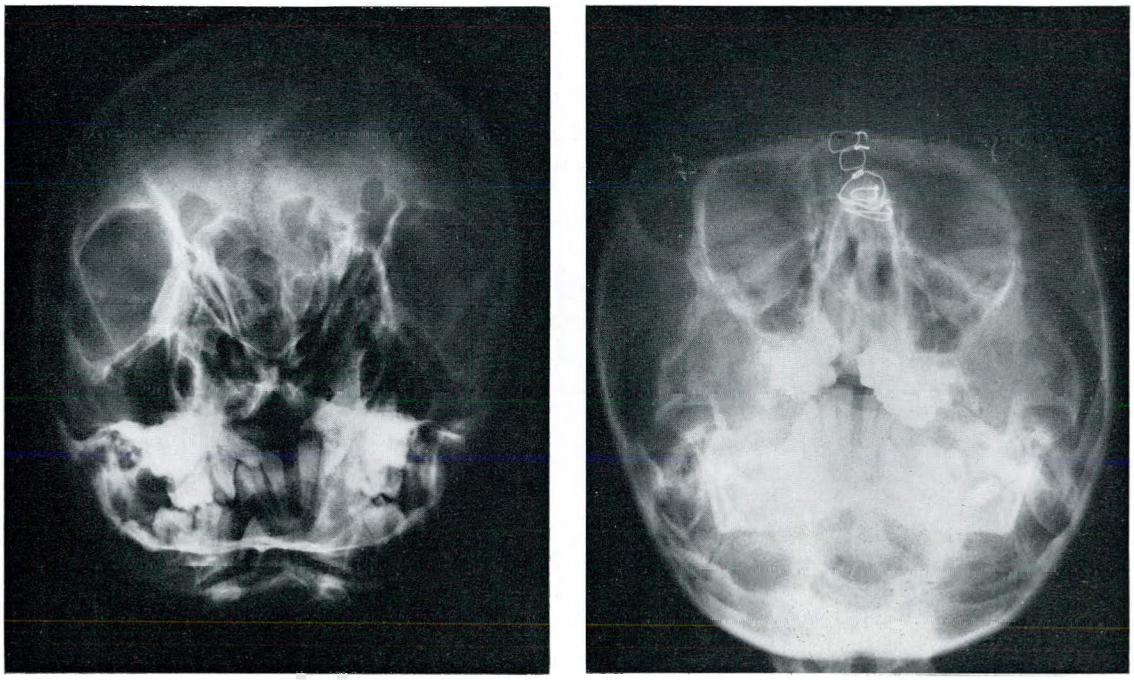


Fig. 10a

Fig. 10b

Fig. 10a + b Radiographs of the skull before and two months after medial rotation of the two facial halves including the orbits. They demonstrate the wide medial rotation of the orbital coni from their extreme lateral position.

moved from the craniotomy sire. The dura was sutured to the replaced bone flap of the skull. A medial canthopexy was performed on each side and the periorbita was fixed to the supraorbital rim. The infraorbital incisions were closed.

After a block of bone was wedged between the rotated maxilla and the pterygoid process on each side, the oral vestibular incision was sutured. No intermaxillary fixation was used because of possible postoperative breathing problems without a tracheostomy. The maxillae seemed stable enough. For the medial rotation of both facial halves the hard palate was reopened. It was then closed, including the cleft uvula. The soft tissues of both nasal halves were then united after excision of surplus skin. The Abbé-flap in the upper lip was excised and both halves of the upper lip were united.

### Severe Postoperative Complications

Postoperatively we had to deal with several severe complications.

Four days after the operation both orbitae were revised as both eyes were rather tense and there was still no movement present. The pupillary reaction, however, had always remained good. On both sides the periorbita was incised. The lateral orbital wall and the floor of the orbits were fractured. No haematomas were found. Two days later the eyes started to move. The patient could count the doctors' fingers.

In spite of 10 million units of penicillin and 1 g streptomycin intravenously per day since the initial operation, both eyelids started to swell again on the 9th postoperative day. There were no other symptoms of severe local infection. On the 11th day after the initial operation the bone flap of the craniotomy had to be removed. Evidence of severe infection was found: bilaterally in the pterygoid-maxillary fossa and the infra- and supra-temporal fossae as well as in the infra-orbital areas; also along the frontal area in direct contact with the infection in the right temporal area. All infected areas were washed and irrigated

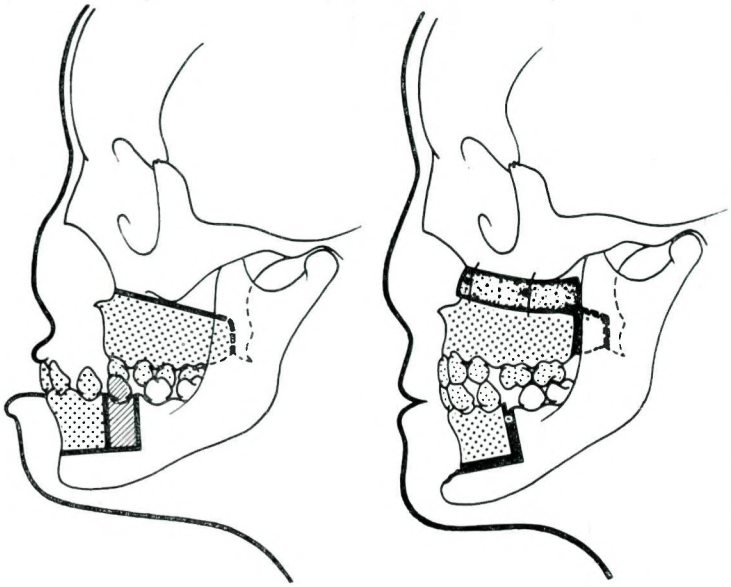


Fig. 11 a

Fig. 11 b



Fig. 12 Two years after primary correction.

Fig. 11 a + b Diagram for the correction of the severe prognathic appearance due to dorsal tilting of the inferior parts of the rotated facial halves due to mandibular alveolar protrusion. Then months after the primary operation the two parts of the maxilla were advanced and the mandibular alveolar segment was retropositioned. a) Indicates the osteotomies on the preoperative skeletal situation. b) Postoperative planning situation with bone grafts in place.

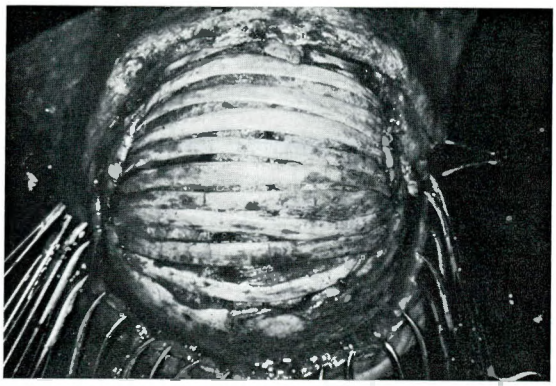


Fig. 13 View of the reconstruction of the large secondary cranial defect by the use of 14 half ribs and deep frozen bank cancellous bone.

with antibiotic solution and drained into the oral cavity. The bone grafts in the lateral orbital regions, in the infra temporal fossae and in the zygomatic arches were removed, since the main abscess formation had developed in these areas.

Two days later bleeding from the right superficial temporal artery caused a huge *subgaleal haematoma*. Again under intubation anaesthesia, evacuation of the haematoma and ligation of a bleeding branch of the right temporal artery was performed.

Twenty days after the initial operation the general condition of the patient became worse although there were no local acute symptoms. Because of his poor general condition the patient was transferred to the intensive care unit of the paediatric hospital. Soon a high temperature developed as well as c. s. f. draining through the vertical medial incision line. The classical symptomatology of a *general septicaemia* developed. Bacterium Coli was cultivated from the blood as well as from the c. s. f. With adequate treatment the temperature became normal after ten days. Four weeks after the beginning of the generalised sepsis the patient was out of bed and one month later he left hospital.

There were four other minor problems which caused us to return the patient to the operating room and have him intubated.

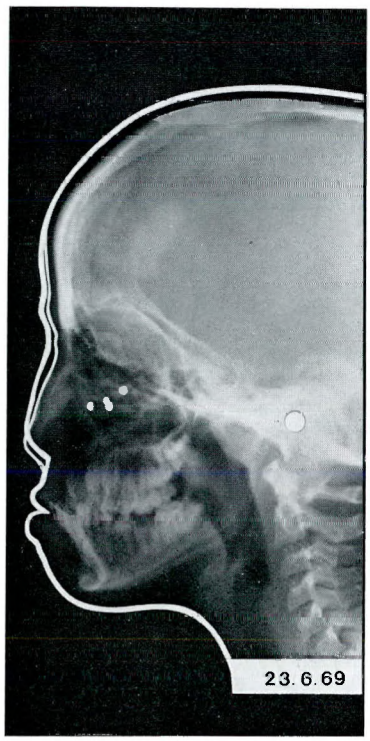



Fig. 14a




Fig. 14b

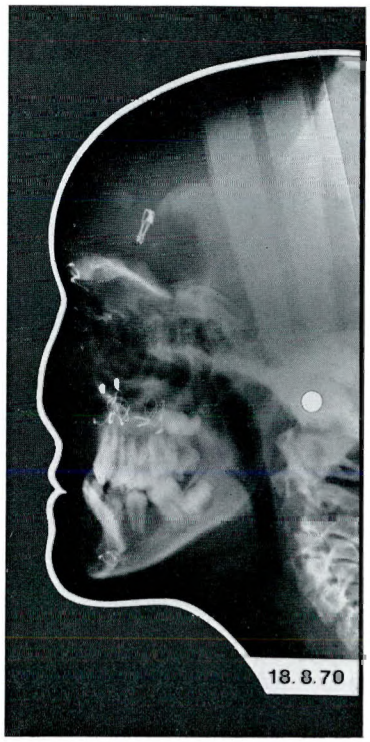



Fig. 14c

Fig. 14a–f Cephalometrics taken during the various stages of the reconstruction: a) Preoperative situation. b) Severe retromaxillism and large cranial defect nine months after primary operation. The latter was due to loss of the cranial bone flap because of infection. The same reason caused the removal of bone grafts behind the medially rotated facial halves which resulted in their backward tilting. c) Intermaxillary and interskeletal relationship four months after advancement of the maxilla and repositioning of the anterior alveolar segment of the mandible. d) Condition of the skull one month after reconstruction of the large defect with 14 halves of ribs. Prognathic appearance becomes pronounced again. e) The halved ribs have become united between each other in spite of the gaps left between them during reconstruction. The middle third of the facial skeleton and in particular the maxilla has not grown forward. A monstrous gorilla-like prognathic appearance has developed between the age of eleven and seventeen. f) Normal intermaxillary and interskeletal relationship after correction of the appearance shown in figs. 14e and 15a + b with the procedure shown in figs. 16a + b.

**Correction of Retromaxillism and Mandibular Alveolar Protrusion**

Seven months after the initial hospital discharge the patient came for follow up. There had been no complications or problems since he had left hospital for home. However, it was obvious that there was a severe retromaxillism and mandibular alveolar protrusion (see *Fig. 14b*). The two halves of the facial skeleton had tilted backwards thus producing a highly prognathic appearance. The tendency to such a change in the position of the maxilla had already become visible before the patient left hospital three months after the facial rotation. The correction of this maxillo-mandibular discrepancy was planned as in any other case of orthognathic surgery *(Fig. 11a+b)*. Ten months after the initial operation, on April 22, 1970 the two halves of the maxilla were advanced by a Le Fort I-osteotomy and the anterior mandibular alveolar segment was retropositioned. Bank bone was used to fill the steps in the anterior maxillary wall. This bone had been taken from the patient during the first operation. An arch bar stabilized both maxillary halves in the transverse plane. Direct bone wires in the canine fossa secured their vertical position. An arch bar fixed to orthodontic bands secured the new position of the retropositioned anterior alveolar segment of the

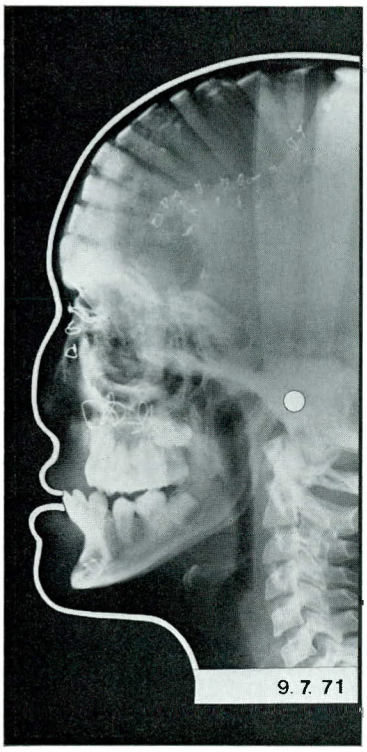



Fig. 14 d

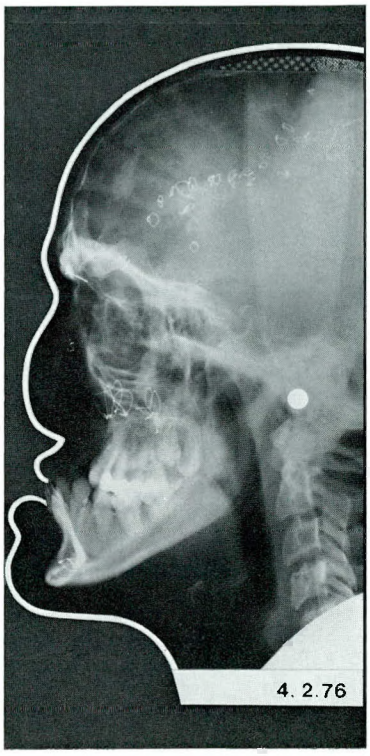



Fig. 14 e

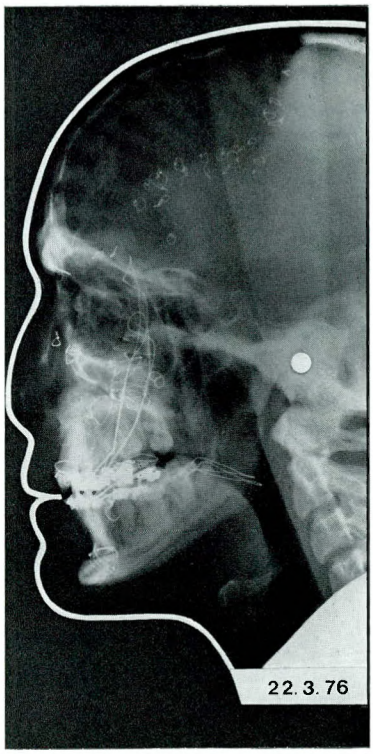



Fig. 14 f

mandible. No intermaxillary fixation was applied. A very satisfactory interskeletal relationship was achieved (see *Fig. 14 c*).

### Replantation of Patient's Cranial Bone Flap

When the patient was discharged from hospital three months after the primary operation he had to wear a helmet because of the large cranial defect. Thirteen months after the bone flap was removed because of infection, it was reimplanted (on August 20, 1970). It had been stored after boiling, following its removal. Because of further infection the bone flap had to be removed again. It was necessary therefore, to reconstruct the cranial defect either with an alloplastic material or with the patients ribs. For this operation an interval of some months was unavoidable. The boy had to wear his helmet again. In spite of the existing defects of the skull and lateral orbital rim areas and in spite of the prognathic appearance increasing once more, the boy had now acquired a rather better looking face *(Fig. 12)*.

### Reconstruction of Cranial and Orbital Defects

Ten months after the unsuccessful attempt at reimplantation of the cranial bone flap the boy returned for a new attempt to have his cranial defect reconstructed.

Within the previous year the patient had grown quite remarkably. It seemed that his skull had become wider in circumference. The hair had fully grown again. The mandible had definitely grown forward. A prognathic intermaxillary relationship and a slightly prognathic appearance had developed again. There was free movement of the eyes. The infraorbital and supraorbital scars were wide and pinkish. The defect of the skull was unchanged. There was no spontaneous bone formation. The areas dorsal to the lateral orbital rims were depressed.

From the right thoracic cage 4 ribs (8th, 7th, 6th, 4th) and from the left 3 ribs (8th, 7th, 5th) were removed. (During the initial operation 3 ribs on the right side had already been removed. They had obviously regenerated but were thinner and had very little cancellous bone.)



Fig. 15 a

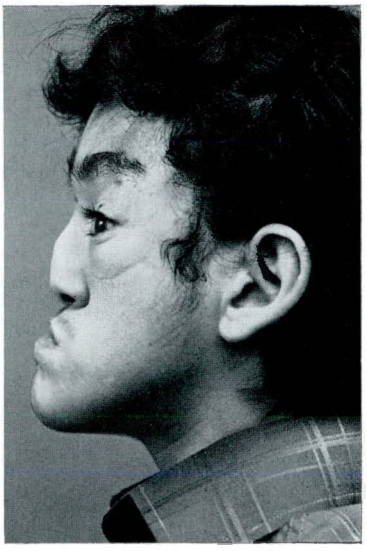


Fig. 15 b

Fig. 15 a + b
Front and side view of the patient's facial appearance when growth had ceased. The monstrous gorilla-like appearance is due to lack of forward growth of the maxilla and zygomatic bones and lower half of nose aggravated by overgrowth of the mandible. They correspond to the cephalometrics shown in fig. 14 e.

On June 9, 1971 the scalp was dissected from the dura. The 14 halves of ribs were used to bridge the defect of the cranium *(Fig. 13)*. The halved ribs were pressed and molded with contouring forceps in order to make them conform with the required curvature of the skull. They were fixed by wires to the free borders of the cranial defect in an inlay-technique manner. Bank cancellous bone paste was used to fill the small gaps between the halved ribs. Sections of halved ribs were used to reconstruct the defects in the lateral orbital walls on both sides.

Because seven ribs had been removed and the possibility of recurrence of severe infection, the boy was taken to the childrens' hospital soon after the operation for the best possible medical care. On the 4th postoperative evening, oedema of the left upper eyelid and left forehead region developed in spite of prophylactic administration of antibiotics. 24 hours later the scalp was raised about 6 cm in diameter by an abscess over the left forehead area. It was evacuated through a stab incision in the eyebrow. It was irrigated twice daily with an antibiotic solution. The halved ribs could be felt with a probe. The feared osteomyelitic destruction of the rib grafts did not occur. Gradually the scalp again became adherent to them. In spite of the removal of seven ribs there were no postoperative respiratory problems.

**Patient's Facial Growth**

The patient's general growth has proceeded normally and increased remarkably between the first operation in July 1969 and February 1976. The shy little boy of ten has become a rather tall young man by the age of seventeen. His facial growth has unfortunately not behaved in the same way over the last seven years *(Fig. 14 a–f)*. The intercanthal distance has increased to 52 mm and the width of the nasal framework has reached 40 mm. These measurements were almost normal at the end of the first operation (see *Figs. 8 and 10 b*). The reconstruction of the cranial defect has become very solid and superficially even (see *Figs. 14 e–f and Fig. 22 b*). As there is no cartilagenous nasal septum there has also been no growth of the lower half of the nose. While the mandible had grown forward tremendously and the area of the frontal bone also seemed to have developed normally, the middle third of the face remained retroposed causing a gorilla-like appearance *(Fig. 14 e and Figs. 15 a + b)*. Detailed figures concerning the boy's facial growth gained from the various cephalometric examinations will be published separately if those taken early on are good enough to show the desired details precisely.

The orthodontic treatment planned initially no longer made sense because of the severe intermaxillary discrepancy. As a result of this gross

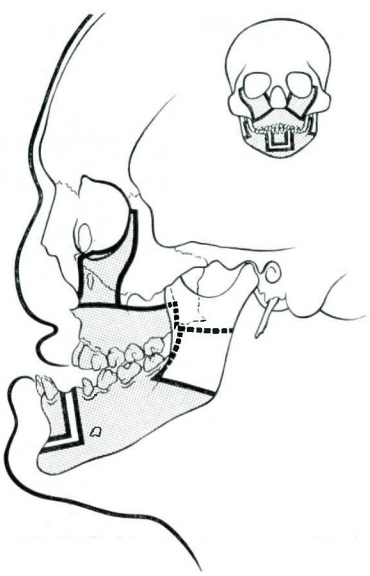


Fig. 16a

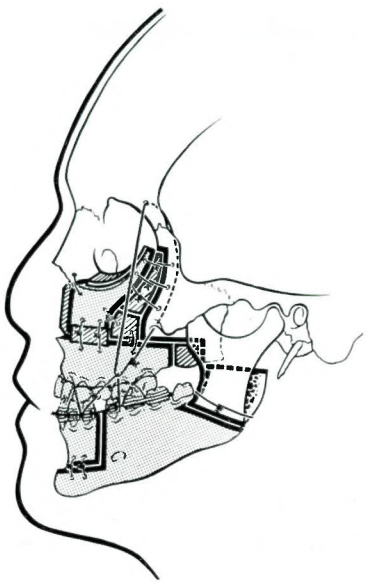


Fig. 16b

Fig. 16a + b
Drawing of the procedure used to correct the monstrous prognathic appearance of figs. 15a + b and 14.
a) The sections of the middle third of the face and of the mandible to be advanced and retropositioned respectively are dotted. The planned osteotomy lines are indicated.
b) Planned situation of the facial skeleton at the end of the correction. Bone grafts are shown by parallel lines in the zygomatic bone and retro-maxillary and canine fossa and orbital floor areas. Interskeletal suspension fixed to the inter-maxillary fixation guarantees the stabilization of the repositioned facial bones.

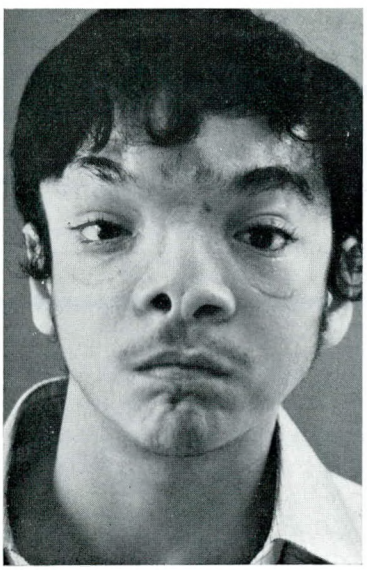


Fig. 17a



Fig. 17b

Fig. 17a + b
Patient's facial appearance five months after the intervention demonstrated in fig. 16a + b.

growth discrepancy in the cranio-facial area another severe cranio-facial deformity had to be corrected. This time the discrepancy was in the anterior-posterior direction while inistially the abnormality had been in the horizontal plane.

**Correction of Gorilla-like Appearance**

After evaluation of the patient's face the following plan was proposed for the correction of the gorilla-like appearance: advancement of the maxilla and increase of the height of the middle

Fig. 18
The first caterpillar flap procedure for elongation of columella and nasal alae is outlined.

Fig. 19
Insertion of L-shaped autogenous cartilage graft for reconstruction of the missing nasal septum and nasal tip and nasal dorsum. Simultaneously a caterpillar flap procedure from the nasal dorsum is again used to reconstruct the missing soft tissues in the nasal tip area.

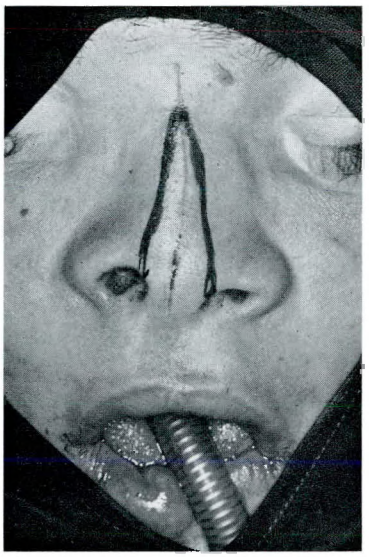


Fig. 18

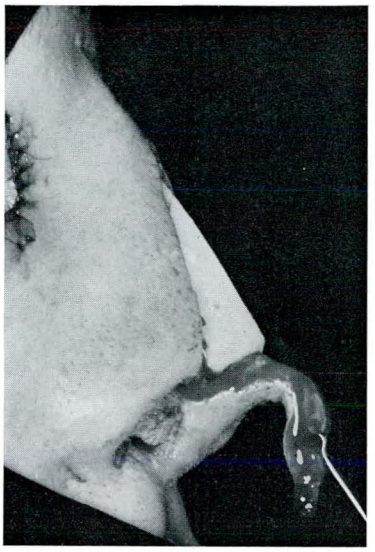


Fig. 19

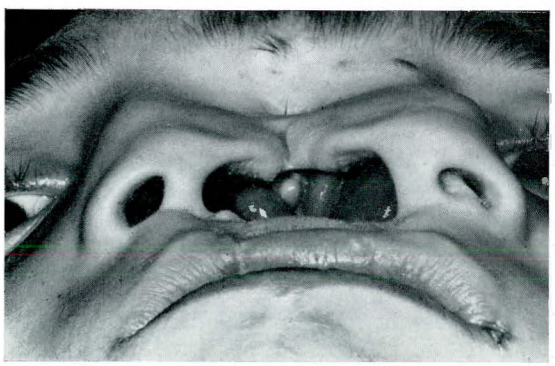


Fig. 20 a



Fig. 20 b

Fig. 20 a + b Close up views from below into patient's nostrils before correction (a) and at the end of corrective surgery (b), eight and half a year after (a). Philtrum and cupid sbow are almost normal.

third; independently the upper half of the middle third had to be advanced but with the exception of the bony nasal framework; the mandible had to be repositioned as a whole as well as the anterior alveolar segment in addition. This skeletal correction was to be achieved in one operation *(Fig. 16 a+b)*. The lack of growth of the lower half of the nose could be dealt with later. On the 6th February 1976 the maxilla was moved forward and downward by a Le Fort I-osteotomy procedure and independently the infraorbital rims with the canine fossae and the medial halves of the lateral orbital rims were advanced on both sides. Only the bony nasal framework and the medial orbital walls with the lacrimal systems were left in place. The mandible was retropositioned using the sagittal splitting procedure and also the anterior mandibular alveolar segment was retropositioned in addition. All defects of bone were filled with bone transplants from the left iliac crest. Intermaxillary fixation ensured the planned occlusion and interskeletal suspension to the supraorbital rims was achieved (see *Fig. 14 f*). After six weeks the interskeletal suspension and the intermaxillary fixation were removed.



Fig. 21 a



Fig. 21 b

Fig. 21 a + b View of the maxilla before and after surgery: a) An abnormally wide upper arch with a large defect in the midline of the alveolus and anterior third of the maxilla and with supernumerary, fully developed front teeth on each half and very poor oral hygiene. b) Almost normal but short upper arch with both canines contacting in the midline. All defects closed.

As a result of this surgery on the middle and lower third of the facial skeleton, a normal intermaxillary relationship and an almost normal facial contour with the exception of the depressed lower half of the nose resulted *(Fig. 17 a + b)*.

**Reconstruction of the Lower Half of Nose and Additional Minor Reconstructions**

Between July 1976 and February 1978 minor surgical procedures were performed in order to complete the case: The broad atrophic infraorbital scars and the hypertrophic scar on the lower lip were excised and closed with an intracutaneous suture which was left in place for over 2 weeks. In spite of this the scars became wide, atrophic and reddish again.

In 3 operations, at intervals of some months the nasal reconstruction was achieved. The lack of soft tissues in the length of the nose as well as in the height of the columella and in the nasal tip and alar areas was overcome by a repeated caterpillar flap procedure from the nasal dorsum *(Fig. 18)*. The missing nasal framework was reconstructed by the implantation of an L-shaped autogenous cartilage graft *(Fig. 19)* and finally the columella was thinned, whereby a very acceptable nasal shape was produced *(Fig. 20 a + b)*. A pharyngo-plasty procedure was performed utilizing a broad cranially based pharyngeal flap. It increased the quality of speech remarkably.

A dacryo-cysto-rhinostomy was performed on the right side. It had become necessary following the advancement of the maxilla and orbital rims and antral walls as a chronic dacryo-cystitis with epiphora had resulted. Drainage remained satisfactory thereafter.

Both doctor or patient might feel that some further minor additional correction is desirable, but a line has to be drawn somewhere.

**Ophthalmological Findings and Evolution**

Originally visual acuity had already been reduced to 0,5 on the left and 0,6 on the right side. The interpupillary distance was increased to 85 mm, compared with a normal of 56 mm. Manifest divergence and fixation nystagmus were observed which were more pronounced on the left than on the right. Otherwise the eyes were normal.

After medial rotation of the orbits two small erosions developed on the right cornea, necessitating partial blepharoplasty. Two months after the operation, visual acuity was 0.4 on the left and only 0.1 on the right. Functional weakness of the right upper eyelid and the external rectus muscle were observed, but the general fixation of the globes caused by postoperative oedema had resolved.

During the years following, visual acuity improved still further and attained 0.4 on both eyes. Binocular vision is now possible for greater distance.

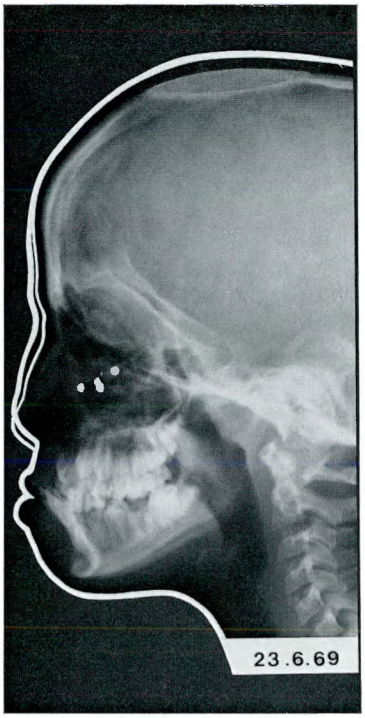



Fig. 22 a

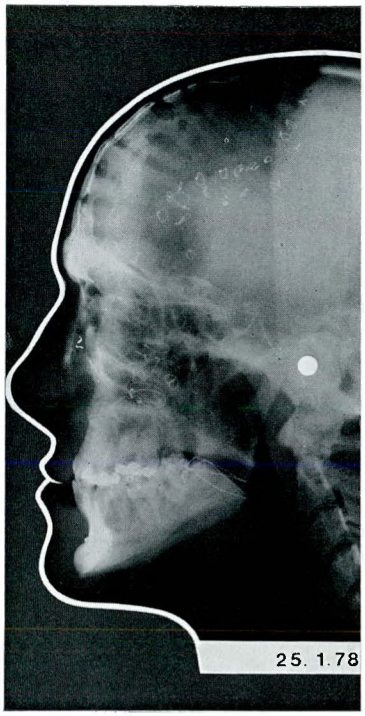



Fig. 22 b

Fig. 22 a + b
Cephalometrics before and at the end of surgery.

a) Demonstrating the two nasal contours and alveolar prognathic appearance.

b) Demonstrating normal profile and skull contours one year and eleven months after last correction of intermaxillary relationship and at the end of reconstruction of underdeveloped lower half of nose. It shows the perfect repair of the skull six years and seven months after its reconstruction with 14 halves of ribs.

but it is impossible for near vision due to lack of convergence. The interpupillary distance measures 67 mm. Slight divergence and vertical strabismus is present, nystagmus is latent. The bulbi are otherwise normal on both sides.

**ENT-Findings**

Preoperatively there were no pathological findings in the external ears, throat or larynx. The abnormalities of the nose were described above. The nasal airways were free. The right tympanic membrane was very thin, retracted and only partially mobile; the left tympanic membrane was not mobile and a thin layer of fluid was visible deep to it. Corresponding to the latter findings the audiogram showed slightly reduced sound transmission. These findings were due to impairment of function of the Eustachian tube, probably caused by insufficient mobility of the soft palate in connection with the operated cleft situation.

Six years after the primary facial rotation the otological findings showed a lateralisation in the *Weber*-test to the right side and in the audiogram a normal threshold of audibility, but on the right a slightly impaired sound transmission.

**Logopaedic Findings**

Before and after the first operation it was almost impossible for Antonio to make himself understood. The vowel sounds were nasal, the consonants uncertain and the articulation not intelligible.

After correction of the gorilla-like appearance and additional velo-pharyngoplasty Antonio could make himself understood, although a mixed rhinolalia and multiple stammering was diagnosed by the speech-therapist.

On the whole, a remarkable improvement of the boy's speech was achieved by surgery alone. Unfortunately it has been impossible to supply the boy with the very necessary speech therapy.

**Result**

An almost normal facial contour and good facial profile have been achieved. There is good intermaxillary relationship with acceptable chewing function, in spite of the fact that all upper incisors are missing. Both halves of the maxilla are properly approximated, forming an acceptable maxillary dental arch *(Figs. 21 a+b)*. The large

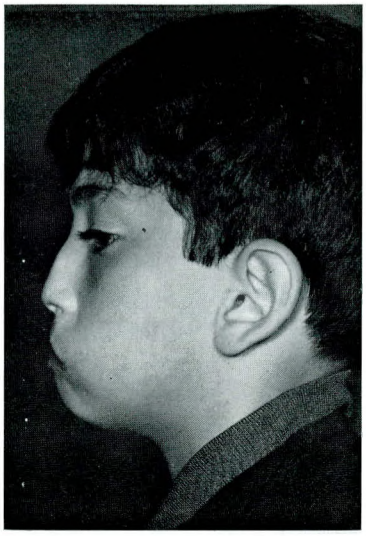


Fig. 23 a



Fig. 23 b

Fig. 23 a–f
Patient's full face and profile appearance and views from before and at the end of corrective surgery: The preoperative photographs are equivalent to the cephalometric view shown in fig. 22 a; the postoperative photographs are equivalent to the cephalometric picture shown in fig. 22 b.

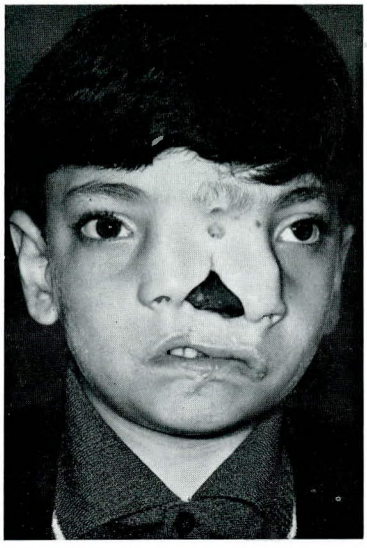


Fig. 23 c



Fig. 23 d

gap in the anterior third of the hard palate and between the two alveolar processes has been eliminated. There is bony union between them giving good support to the L-shaped cartilage resting on it. The final result of the patients facial appearance is demonstrated in *Figs. 22 and 23.*

The nasal airway is free and there are no symptomatology and complaints in spite of the missing nasal septum. The speech quality has improved very remarkably. The open nasality was reduced following pharyngoplasty. However, there was still some degree of open nasality. As a whole the speech quality is quite satisfactory. The patient can be understood easily. Formerly this was not the case.

There is no evidence of any severe, permanent damage to the eyes. While immediately after the operation the condition of the eyes was considered alarming, the later follow-ups showed that most of these adverse findings had disappeared

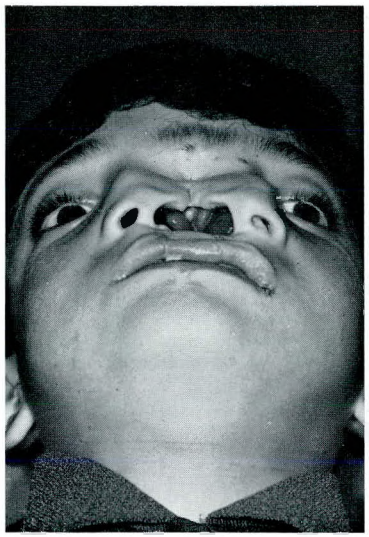


Fig. 23 e

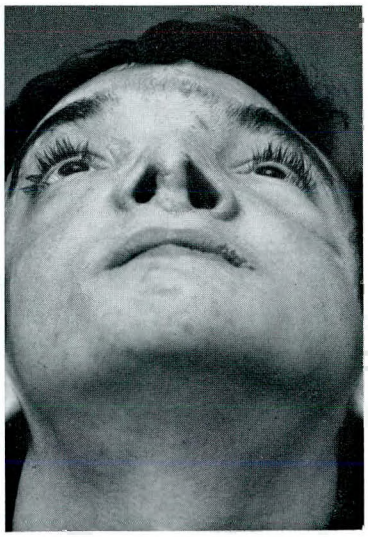


Fig. 23 f

and that no harm was done to the eyes except a slight reduction of visual acuity; this however, was not appreciated by the patient.

From the otological point of view there was no improvement of function of the Eustachian tubes after advancement of the midface and velopharyngoplasty; the slightly reduced sound transmission already present preoperatively, remained, but was not at all relevant. The patient's sense of taste remained normal, but there was no sense of smell on completion of the surgery. The preoperative hemispastic syndrome was uncharged. There was no other additional neurological symptomatology.

The main problem from the surgical point of view still remaining is the poor quality of the scars in both infraorbital regions and in the lower lip. The scars were wide, atrophic and reddish in spite of the fact that they had been carefully corrected 3 times, even with intracutaneous stitches left in place for more than 2 weeks. There was the same poor quality of scars in the chest and hip areas where the bone grafts had been taken. Only on the dorsum of the nose and in the columella were the scars almost invisible.

## Discussion and Conclusion

There are not many cases with an almost complete duplication of the medial section of the middle and upper thirds of the face published in the literature.

Certain milder degrees of midfacial duplication are also published under the terms "Median cleft face syndrome" (*DeMyer* 1967), "Midfacial clefts" (*Iregbulem* 1978). *Potter* and *Craig* (1976) reproduced the pictures of two cases with complete or almost complete nasal duplication and *Millard* (1977) reproduces a photograph of a case who seems to be a mirror-image of our case when he was a baby. There was even an extra midline orbit together with its eyebrow as in our case.

The authors mentioned above, as well as others (*Goulian* and *Conway* 1964), have published some early results of their surgical correction of their midfacial cleft cases. Unfortunately there is no report of a single case which has reached its final facial growth after the facial duplication has been treated in childhood. It seems that our case is the first from which a lot can be learned about the behaviour of the skeletal structures of the middle and upper thirds of the cranio-facial skeleton following surgery in early childhood. We have published this case previously, but only with the immediate result of the rotation of his facial halves (*Obwegeser* 1974, 1976) and with the change of his profile during growth (*Obwegeser* and *Freihofer* 1977). It is of course known that there is a remarkable growth disturbance

after advancement of the maxilla or of the whole middle third of the facial skeleton if these operations are performed during the growth period (*Freihofer* 1973, 1976, 1977, *Hogeman* and *Wilmar* 1974, *Obwegeser* and *Freihofer* 1977).

Although nowadays it seems to be very common for severe hypertelorism to be corrected in early childhood (*Tessier* 1971, *Edgerton* et al. 1974, *Converse* et al. 1974, 1975) there are as yet no reports concerning the growth behaviour of those sections of the cranio-facial-skeleton which have been involved in this kind of surgery. Therefore it is important that the long term results of every single case, and the experience gained in this respect, is published.

The case published was our first case of transcranial hypertelorism correction. The important principles for correction were given by *Tessier's* ingenious pioneering work on this subject (*Tessier* et al. 1967). It was therefore only necessary to evaluate the anatomical details of our case and accordingly to plan the required surgery. It has proved very useful to have a written plan of the operation laid down in a step by step manner, and given to everybody involved with the operation, including the observers. Since this was so useful in the first case we have also done it in every subsequent case. It has another advantage: it causes the principal surgeon to take the necessary time to sit down and go through the problems of the case in detail and maybe even discuss some of the steps with his neurosurgeon, anaesthetist, co-workers, or the principal nurse. It makes things much easier during surgery and we recommend it highly.

We have never found tracheostomy for intubation anaesthesia necessary for performing major cranio-facial surgery and this is since 1967, soon after *Tessier's* first publication (*Tessier* 1967, *Tessier* et al. 1967). In this case we used oral intubation. To a certain extent this was a disadvantage at the end of our operation; we could not apply intermaxillary fixation. However, it was not planned primarily. Our very experienced anesthetist brought the case nicely through the long operation in spite of the fact that in those days we did not have a monitoring-system for the patient's blood pressure, temperature, electrocardiogram etc. Soon after the last stitch uniting both halves of the upper lip the boy started moving and a few minutes later he was opening his eyes.

One of the problems we had to deal with several times during the first postoperative days was bleeding. There was not much bleeding during the operation; 6700 ml (ccm) of blood were used. This, of course, is much more than his own whole blood volume. At the end of operation when we closed the scalp incision there was bleeding from even very small vessels. Since then we no longer use a large quantity of citrated blood, but always a certain amount of fresh blood.

The local infection in the retromaxillary and infratemporal spaces did a lot of harm, as consequently we had to remove the bone grafts in these areas. Nowadays we are of the opinion that it is not necessary and even wrong to remove the bone grafts because of an acute infection. Nowadays in all our cases of midfacial or cranio-facial surgery the retromaxillary and infratemporal spaces are drained into the oral cavity. In spite of this we have had no problem with a possible ascending infection or with the loss of the bone grafts because of osteomyelitis in these areas. Of course, vacuum-drains are placed between the skull und scalp. – Particularly impressive was the fact that the acute and massive infection in the left supraorbital area after the reconstruction of the large cranial defect by the use of 14 halves of ribs did not lead to any loss of these ribs. Because of the abscess formation in that area the scalp was elevated from the ribs over an area of about 6 cm diameter. There was plenty of foul smelling pus. The abscess was evacuated through an incision in the eyebrow and the abscess cavity was washed out and irrigated twice daily with an antibiotic solution. Within 10 days the scalp was again adherent to all the underlying bone.

The postoperative retro-displacement of the inferior portion of the upper two thirds of the facial skeleton could have been avoided if we had not removed the bone grafts in the lateral orbital walls and the infra-temporal and zygomatic arch areas. Obviously the pterygoid processes were fractured. The bone grafts wedged between the tuberosities of the maxillae and the pterygoid processes, could not therefore prevent the retro-displacement. The existence of the retro-displacement of the maxilla induced us to do in additional Le Fort I-osteotomy advancement at the

age of 11. Nowadays we know by our late follow-up results (*Freihofer* 1977) that this is a severe insult to the growth of the maxilla. It was therefore an additional cause for the subsequent development of the gorilla-like appearance which was also due to the overgrowth of the mandible and the flatness of the lower half of the nose.

While we noted a tremendous change in the intermaxillary relationship during the growth period, there was practically no change after the correction of the gorilla-like appearance which was performed after facial growth had ceased. After the latter procedure only a slight forward tilting of the mandibular anterior teeth occured.

Because of the gross influence of our surgery on the growth of the middle third of the facial skeleton we suggest that whenever possible surgical correction be postponed until growth has ceased. In this specific case we would definitely have saved the patient a great deal of secondary surgery.

The lack of a nasal cartilaginous septum following the initial corrective surgery at the age of ten resulted in a remarkable lack of growth of the soft tissues of the lower half of the nose. It influenced adversely the growth of the columella as well as of the alae, in spite of the fact that the alar and the triangular cartilages were present. We would, therefore, if ever we have to correct a case with two nasal septi again, try to save one of them.

The vertical scar in the midline of the forehead and the scars on the nose after multiple corrections have become almost invisible. However, the infraorbital scars and the scar on the lower lip after an Abbé-flap operation in early childhood, remain very unsatisfactory in their appearance, despite multiple corrections. Whenever possible, our policy has been one of avoiding incisions in visible areas and thereby preventing unsightly scars after maxillo-facial surgery. We no longer use infraorbital incisions for achieving access to the orbital floor and the infraorbital rim. The subciliary or the transconjunctival approach are much better. However, even these incisions for osteotomies in these areas are not essential. Our temporal approach (*Obwegeser* 1978) permits good access to the orbital coni in osteotomies for Le Fort III-advancement and transcranial correction of hypertelorism. No facial skin incisions are needed.

We observed spontaneous regeneration of the removed ribs if the periosteal tube is carefully sutured. The regenerated ribs, however, are thinner than normal and they contain very little cancellous bone. The subperiosteal removal of seven ribs in one operation, three on the one side and four on the other side, does not cause any severe postoperative respiratory problems.

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But it is especially Antonio himself, who has to be thanked for his inexhaustible confidence in the arts of medicine in spite of the long-lasting treatment and all its complications.

In the same way, we are greatful to Antonio's parents who, like him, have believed in us and therefore have contributed highly to the result which was finally achieved.

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