HARELIP AND CLEFT-PALATE*

A STUDY OF FOUR HUNDRED AND TWENTY-FIVE CONSECUTIVE CASES

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of Philadelphia, Pa.

The four hundred and twenty-five cases in this series include the various types of harelip and cleft-palate deformities which have been under my care from January 1, 1914, to November 1, 1927. I am indebted to Dr. J. Chalmers DaCosta for the opportunity of caring for all harelip and cleft-palate cases admitted to "Surgical Division A" at the Jefferson Hospital since 1915, which cases constitute nearly one-half of this series. The other cases were those on my service at the Philadelphia General Hospital from 1915 to 1920, those at the Frankford Hospital since 1922 (for which I thank Dr. Charles F. Nassau), and my private cases.

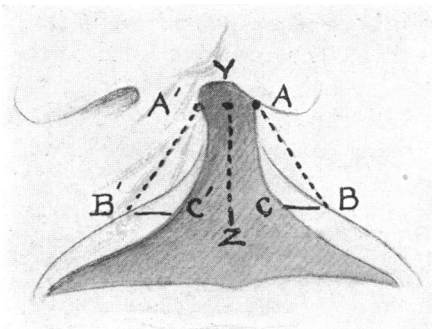

Fig. 1.—Semidiagrammatic sketch, showing lines of incisions used for the correction of unilateral harelip after the method of J. E. Thompson. Sharp pointed calipers are used in measuring the distance YZ from the midpoint of the floor of the nostril being constructed to the point in the same sagittal plane to which the free margin of the lip would come if it were of normal contour. Fixing the distance on the calipers and keeping the superior point at Y, the inferior point of the calipers is rotated describing an arc which crosses the vermilion border of the lip on either side of the cleft. These points B and B′ are distinctly marked by making a puncture with the point of the calipers or with a small scalpel. Points C and C′ are then located on the free margin of the lips so that the angles ABC and A′B′C′ are between 70 and 80 degrees. Incisions carried through the entire thickness of the lip with a small scalpel at a right angle to the skin surface and following the lines as outlined will give surfaces for approximation which are of equal length and which, when sutured together, will give a lip the length of which is the estimated normal length YZ plus the distance from the vermilion border to the free edge of the lip CB which is usually just sufficient to allow for subsequent contraction.

Fifty-six per cent. of the cases were males and forty-four per cent. females. The age of the patients at the time of first examination varied from one day to seventy-six years. Sixty-three per cent. of the cases were under seven months old. The types of the deformities, the number and the percentage of each type are shown in Table I.

Etiologic considerations show that heredity is a dominant factor, positive family histories being obtained in fifty-seven per cent. of our cases. The percentage doubtless would have been considerably greater if the knowledge of the family histories had been more extensive and accurate. In many cases our information was limited to only one or two generations. By positive history we mean not only the occurrence of actual clefts of lip or palate of some degree in some other member or members of the family within three generations, but also the congenital absence of one or both of the permanent superior lateral incisor teeth in some of the relatives. In

* Read before the Philadelphia Academy of Surgery, December 5, 1927.

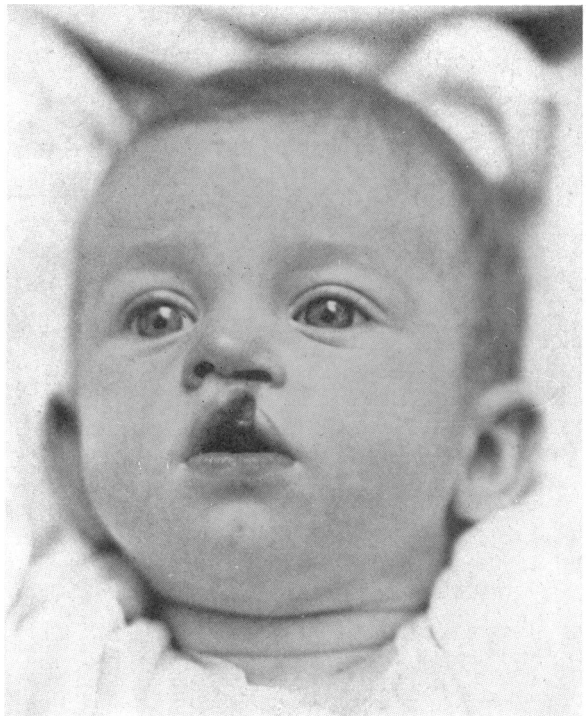

FIG. 2.—Case I. B. W., child, age seven months, with incomplete unilateral harelip. Note the absence of muscle tissue between the superior angle of cleft and floor of the nostril, deviation of the nasal septum, and flattening of the ala.

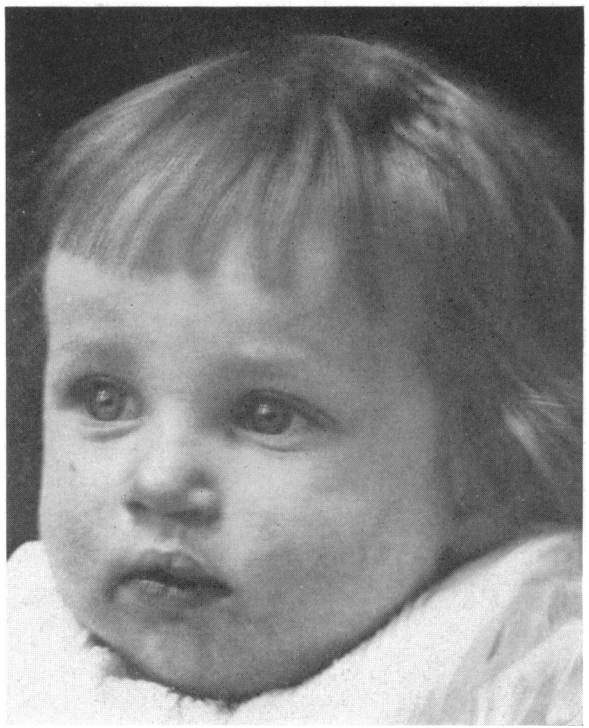

FIG. 3.—Case I. B. W., ten months after operation, showing contour of lip and nostril. The incisions used in this case were those of the Thompson method, carrying the incisions up into the floor of the nostril, removing sufficient tissue to allow the proper approximation of the ala to the septum after freeing the lateral portion of the lip and the antero-inferior portion of the cheek from the maxilla.

eighteen cases we have found this relationship existing between absent lateral incisor teeth in one generation and the presence of harelip or cleft-palate, or both, in a succeeding generation. In four instances the absence of the lateral incisor teeth occurred in a parent, in five instances in an uncle or aunt, and in nine instances in first or second cousins of the patient. We have been greatly interested in this tendency which we believe has not been previously pointed out. In the cases with a negative history for actual clefts we are now requesting the parents to coöperate in searching for relatives showing congenital absence of the lateral incisor teeth. We have not been able to prove syphilis an etiologic factor in any case. Race has some bearing—only one of our cases being a negro. Only six per cent. of the cases were Hebrews.

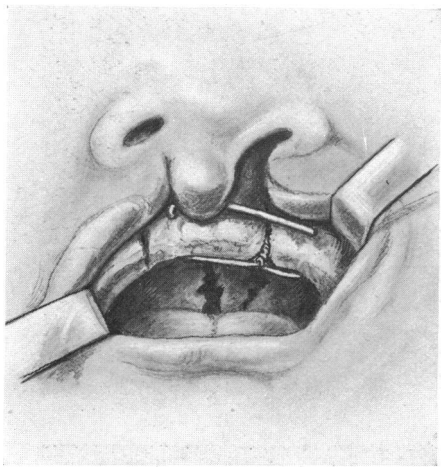

FIG. 4.—Semidiagrammatic sketch showing partial division of alveolar process posterior to the canine area on the right side to facilitate bringing premaxilla into normal position. A wire suture is used to hold the parts in apposition.

Other congenital defects which we found associated with harelip and cleft-palate deformities were supernumerary digits in four cases; deficiency in number of digits, one case; webbed fingers, one case; club-foot, one case; spina bifida, one case; hernia, eight cases; deficient development of mandible associated with microglossia, two cases; mongolian idiocy, one case; idiocy, three cases; distinctly defective mentality of less degree than idiocy, five cases.

The great value of the splendid coöperation which we have had from the Pædiatric Department at Jefferson, and also at the Frankford Hospital, is fully appreciated. Many of the infants were sent to the hospital in a very

TABLE I.

Unilateral harelip (left)	34 cases or	8%
Unilateral harelip (right)	21 cases or	4.94 + %
Bilateral harelip	13 cases or	3.05 + %
Median harelip	2 cases or	0.47 + %
Unilateral harelip and cleft-palate (left)	132 cases or	31.05 + %
Unilateral harelip and cleft-palate (right)	26 cases or	6.11 + %
Unilateral harelip and bilateral cleft-palate	8 cases or	1.88 + %
Bilateral harelip and unilateral cleft-palate	6 cases or	1.41 + %
Bilateral harelip and bilateral cleft-palate	55 cases or	12.94 + %
Unilateral cleft-palate extending into hard palate, without lip deformity	9 cases or	2.11 + %
Bilateral cleft-palate extending into hard palate, without lip deformity	85 cases or	20%
Cleft involving soft palate only	34 cases or	8%

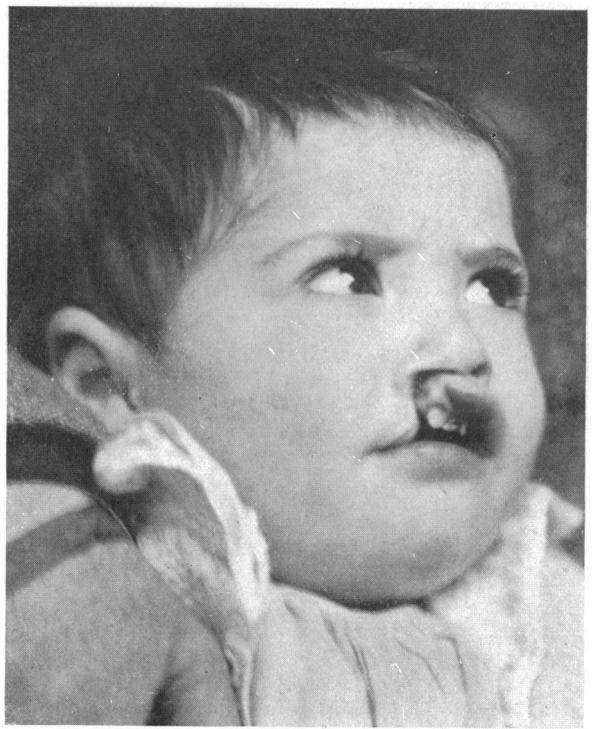

FIG. 5.—Case II. R. M., age seventeen months, showing complete unilateral harelip and cleft-palate. Note rotation of premaxilla and flattening of the nostril.

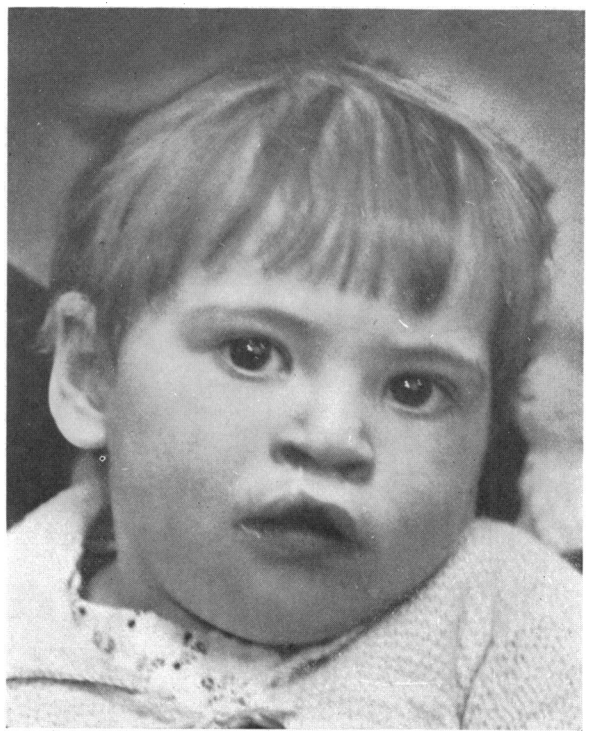

FIG. 6.—Case II. R. M., showing contour of lip and nostril six months after operation. Alveolar process was partially divided lateral to the left canine area to allow the premaxilla to be forced into approximately normal position, as shown in Fig. 4. This procedure also corrected the marked deviation of the nasal septum. Incisions for the lip were outlined by the Thompson method.

FIG. 7.—Case III. E. C., age ten weeks, showing an unusual, wide and extensive unilateral cleft of lip and palate in which the bony cleft extended into the floor of the orbit. The alveolar process was approximated and the lip reconstructed at the first operation.

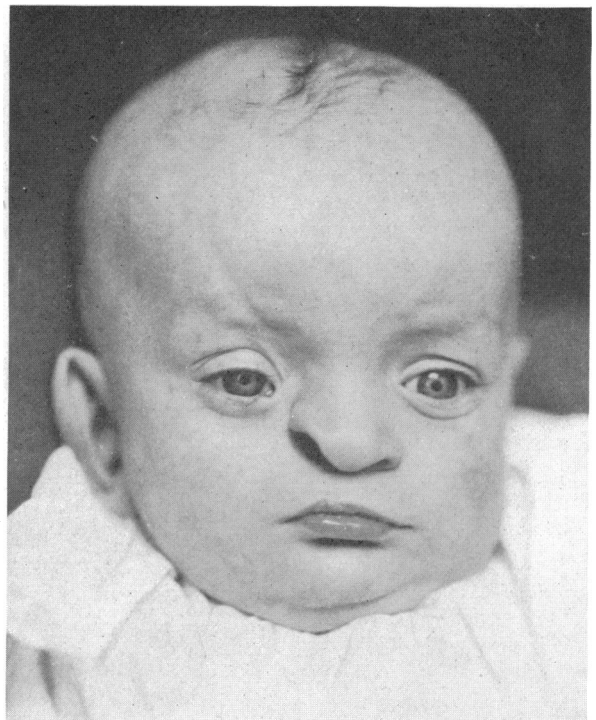

FIG. 8.—Case III. E. C., showing contour of the lip and nostril, eight months after operation. Fourteen months after the first operation a second operation was done to correct the deformity of the right nostril. This was done by detaching the ala laterally and along the mid portion of the right nasal bone allowing the ala to swing into normal position as a pedicled flap where it was sutured with an inner and outer row of interrupted black silk sutures. The area over the nasal bone from which the superior portion of the flap was removed was allowed to heal by granulation. Vaselinized dressings were used. The area healed so rapidly that skin grafting was not deemed necessary.

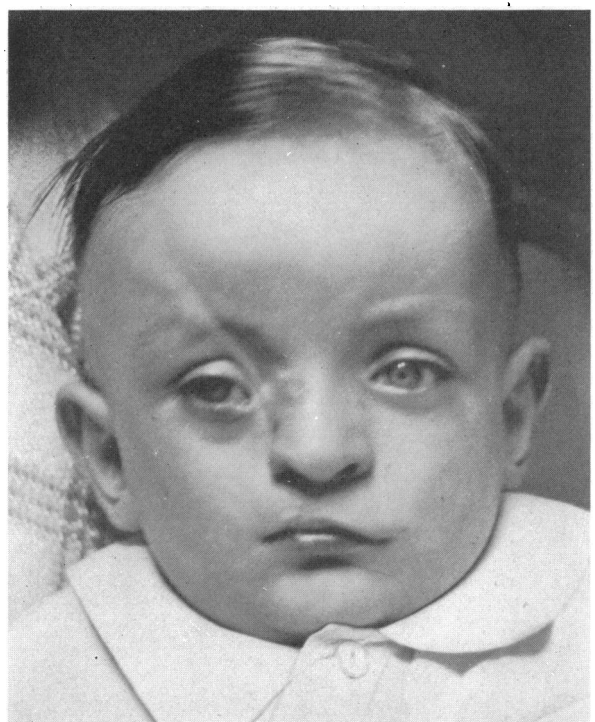

FIG. 9.—Case III. E. C., appearance of lip and nostril five months after second operation.

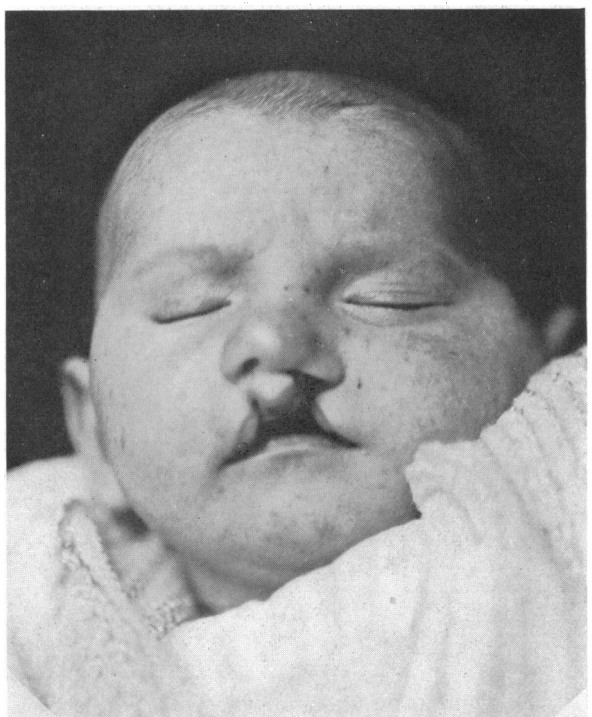

FIG. 10.—Case IV. M. M., age two months, eight days, showing complete unilateral cleft of lip and palate left side, incomplete harelip right side. Alveolar process was partially divided lateral to the right canine area allowing premaxilla to be forced into normal position where it was held with silver wire. The left side of the lip was repaired at the same operation.

FIG. 12.—Case IV. M. M., showing appearance of lip and nostril seven months after second operation. Note that the philtrum has developed until it is now of approximately normal length and the contour of the lip is almost normal.

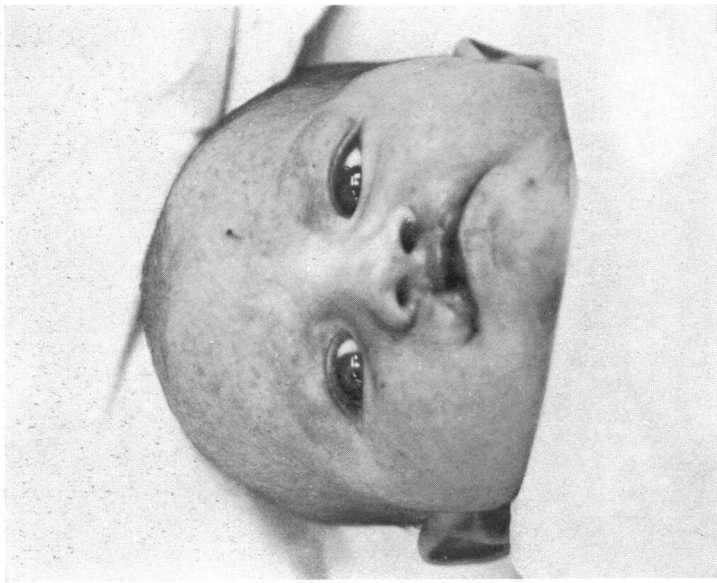

FIG. 11.—Case IV. M. M., showing the condition of the lip and nostril four weeks after the first operation. At the second operation the right side of the lip was repaired. Care was taken in both operations to preserve the entire philtrum. (Fig. 20.)

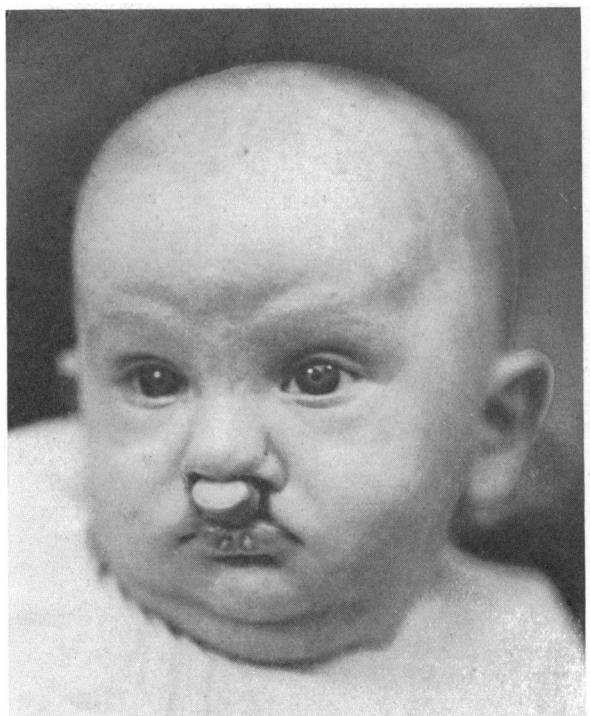

FIG. 13.—Case V. T. H., age five months, complete bilateral harelip and cleft-palate showing moderate anterior rotation of premaxilla. Note the presence of mucous pits on the lower lip which are rather rare, having been present in only three instances in 425 cases. The premaxilla was replaced as outlined in Figs. 18 and 19. The lip was repaired by the method illustrated in Fig. 20.

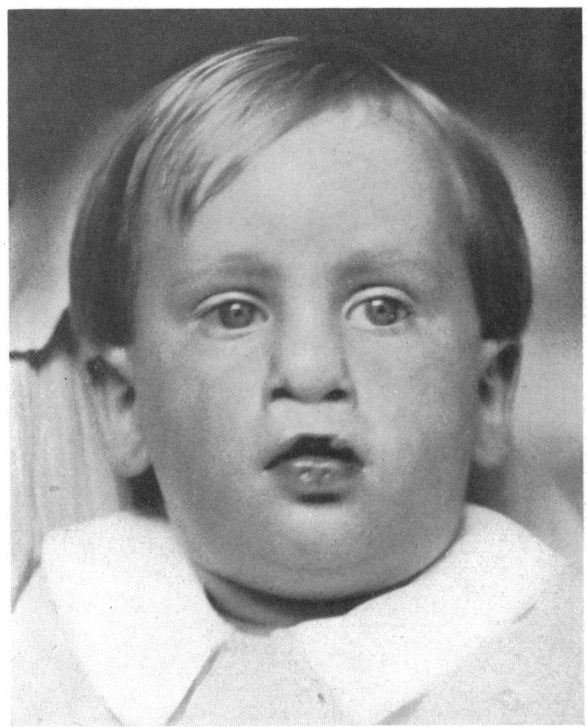

FIG. 14.—Case V. T. H., showing the appearance of lip and nostrils twenty-two months after the first operation. The bilateral cleft-palate was repaired twelve months after the first operation. The slight amount of excess tissue at the free margins of the lip on the left side will be removed later as was done in Case VI, Fig. 16.

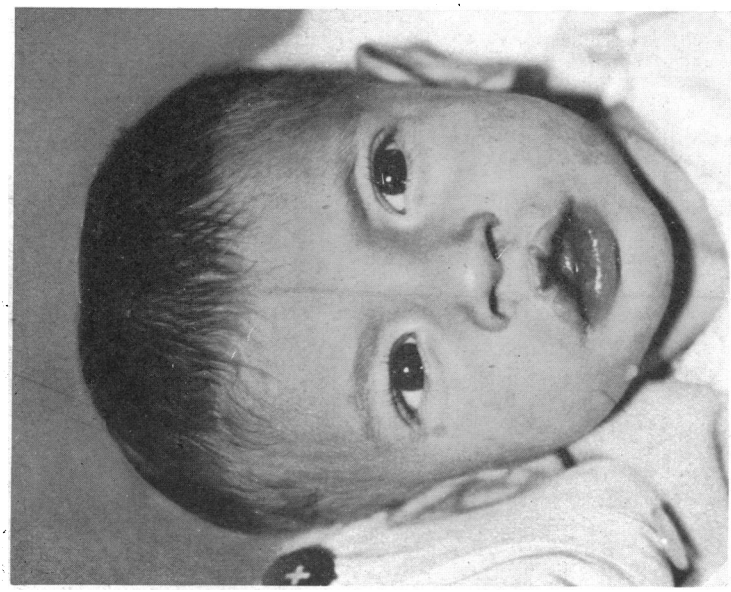

FIG. 16.—Case V. C. G., showing the appearance of lip and nostril five months after operation. The cleft in the palate was closed at this time. Eight months later the excess fullness of the lip lateral to the philtrum was corrected by removing a small amount of tissue from the postero-inferior surface of the lip on each side.

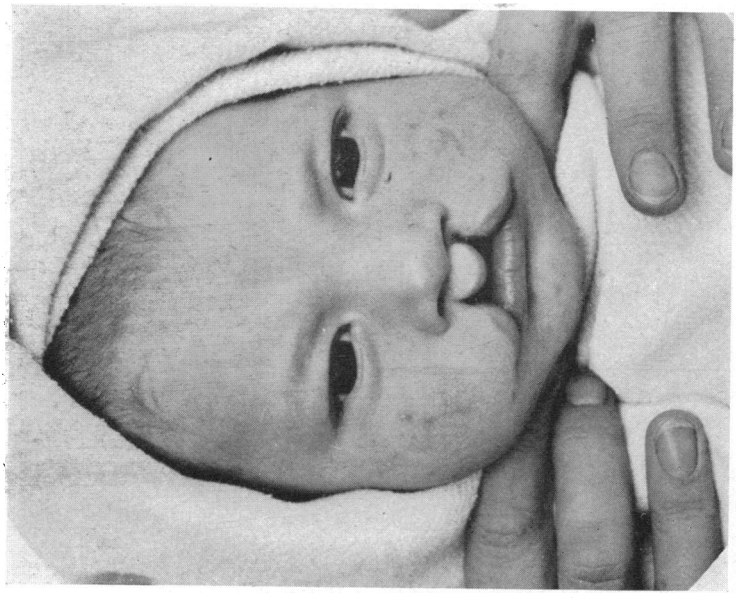

FIG. 15.—Case VI. C. G., age four months, bilateral harelip and cleft-palate. Displacement of the premaxilla was not marked in this case. Nearly the entire philtrum was utilized in the repair of the lip. The incisions were outlined as in Fig. 20.

poor state of nutrition and required much pædiatric care before their general condition would permit operative measures.

In nine cases of bilateral cleft-palate the development of the horizontal processes of the palate was so very rudimentary that operative measures were deemed inadvisable. In such cases dental plates supported by some of the teeth are used. Eleven cases included in this series are not yet in condition for operation. Twelve infants, who were never operated upon, died from nutritional disorders or respiratory infections. Thus three hundred and ninety-three cases have been operated upon, the number of operations required on each varying from one to four, according to the type of the deformity. The surgical mortality was as follows: One case died on the operating table shortly after the administration of ether was begun and before any incision was made. Post-mortem examination showed an enlarged thymus and general lymphatism. Three died within twenty-four hours after operation from shock and pulmonary œdema. Four died after the twenty-four-hour period and within six weeks, from pneumonia. A total of eight deaths or a two per cent. surgical mortality. One infant who died three weeks after operation from impetigo contagiosa bullosa (contracted in the ward one week after operation) is not included as a surgical mortality.

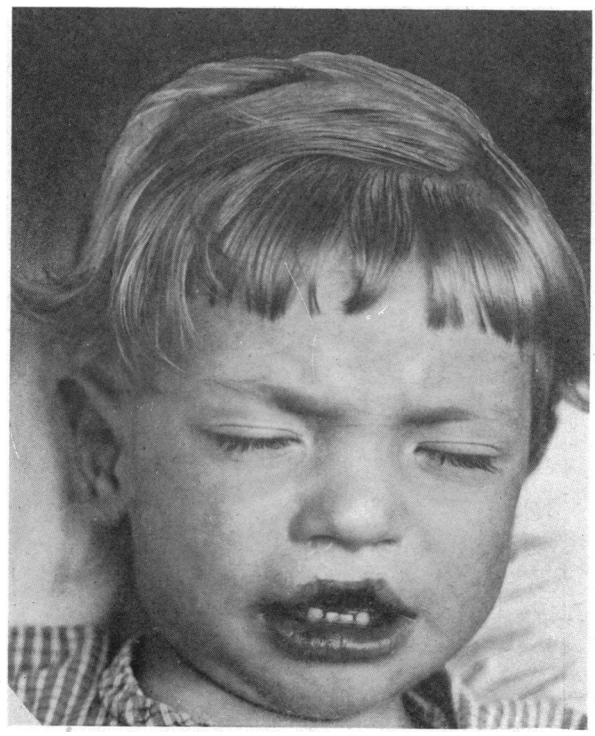

FIG. 17.—Case VI. C. G., showing appearance of lip and nostril fourteen months after the first operation on the lip and one month after second operation.

Careful observation of the ultimate conditions following the different types of operations shows that the best functional and cosmetic results were those obtained by the operative procedures, briefly outlined below, and described in the legends accompanying the illustrations.

Harelip deformities should be corrected as soon as the child is in condition to stand the operation—which is usually sometime between the tenth day and the third month. Incisions are outlined by the Thompson method. (Fig. 1.)

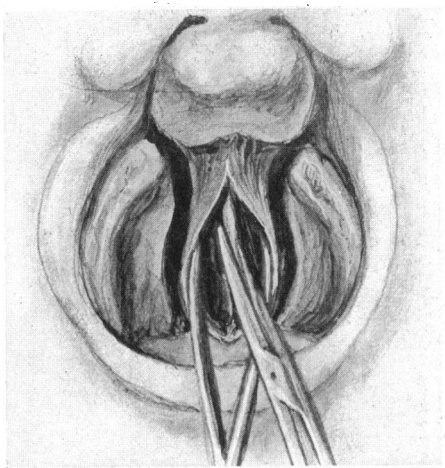

FIG. 18.—Sketch showing method of removing a section from the lower portion of the vomer and the anterior portion of the nasal cartilage. The removal of a triangular section of bone and cartilage allows infero-posterior rotation of the premaxilla to approximately its normal position. The length of the base of the triangular piece of bone and cartilage removed is determined by the amount of rotation which the premaxilla requires, and should be such that when the premaxilla comes into proper position the sides of the triangle will be brought together. There will be bulging of the mucoperiosteum at this point for several days, but the excess tissue soon resorbs.

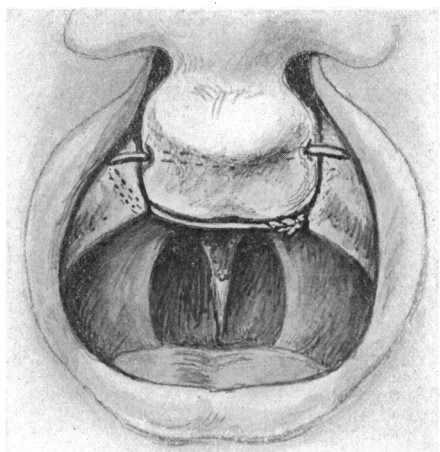

FIG. 19.—Lateral margins of the premaxilla and the margins of the alveolar process have had the mucosa removed from the points which will come in contact to allow approximation of the raw surfaces and permit fibrous union. The premaxilla is held into position by a wire suture which passes through the superior portion of the alveolar process on each side. It is carried in front of the premaxilla but posterior to the philtrum.

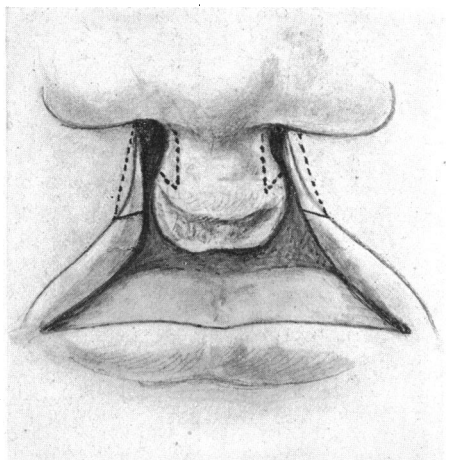

FIG. 20.—Sketch showing the approximate outlines for incisions which allow preservation of almost the entire philtrum in the closure of bilateral harelip. Incisions along these lines are carried through the entire thickness of the lip and the philtrum. The alæ are brought into proper relation to the septum after which the points of the free edges of the philtrum and the corresponding points on the lip are brought into accurate apposition using care that the vermilion borders of the lateral portions of the lip and those of the philtrum are even. The lips shown in Figures 10, 13 and 15 were repaired by this method.

FIG. 21.—Case VII. B. D., age two months. Approximate three-quarter view from the right side showing a very unusual bilateral cleft of the face. The bilateral cleft involved the lip and the alveolar process, but the palate posterior to the premaxilla was united. The clefts in the lip do not extend into the nostrils as in an ordinary case but pass lateralward extending into the orbits. These clefts into the orbits involve the bony structures as well as the soft tissues. Note the extensive coloboma. The naso-lacrimal ducts do not communicate with inferior nasal meatus but pass posterior to the premaxilla and open into the roof of the mouth. Examination of the eye grounds by Doctor Shannon showed the fundus of each eye apparently normal.

FIG. 22.—Case VII. B. D., Three-quarter view from the left side. Note that the opening of the anterior nares is approximately on the same level as the pupils of the eyes. The anterior nares, although small, were well formed. The posterior nares were also well formed, but midway between the anterior and the posterior nares there was complete obstruction on both sides formed by a transverse partition of soft tissue covered by mucous membrane.

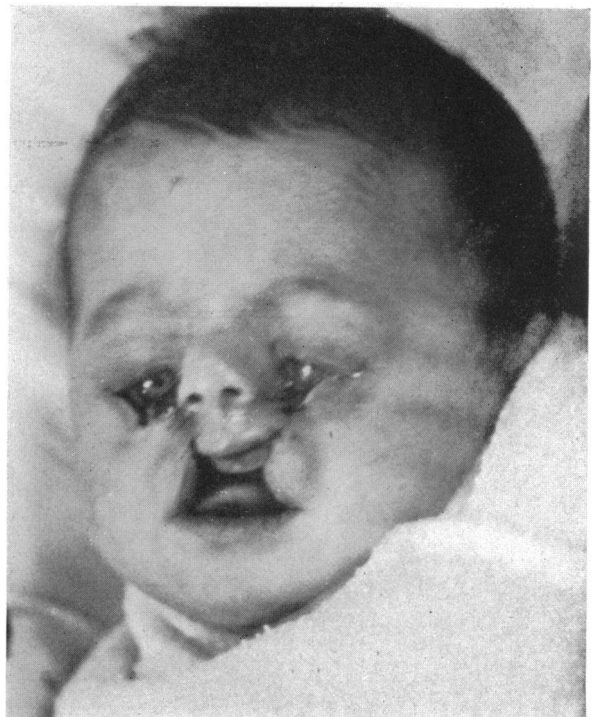

FIG. 23.—Case VII. B. D., one month after the first operation, at which time the premaxilla had been forced into as near the normal position as possible, the cheeks freed from the anterior surfaces of the maxillæ, and brought medially and superiorly to form a floor for the orbit and to bring the lower eyelid medialward. Two operations were done after the stage shown in the above illustration for the further repair of the lip and the eyelids. A fourth operation was done to make an opening through the partition between the anterior and the posterior nares.

FIG. 24.—Case VII. B. D., showing condition of face three months after the first operation. Four operations were performed. The patient is to return in twelve months for further minor corrections.

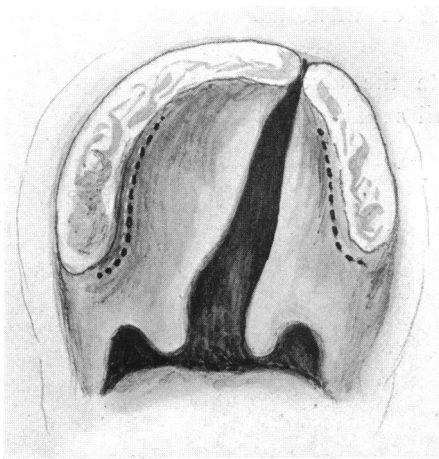

FIG. 25.—Semidiagrammatic sketch showing unilateral cleft-palate and the outline of incisions (slight extension of the Langenbeck incision) used in loosening the mucoperiosteal flap on the side attached to the vomer and loosening the flap containing the rudimentary horizontal process of the maxilla and of palate bone on the opposite side. The latter incision is carried through the periosteum after which the horizontal processes are cut through with a very thin chisel.

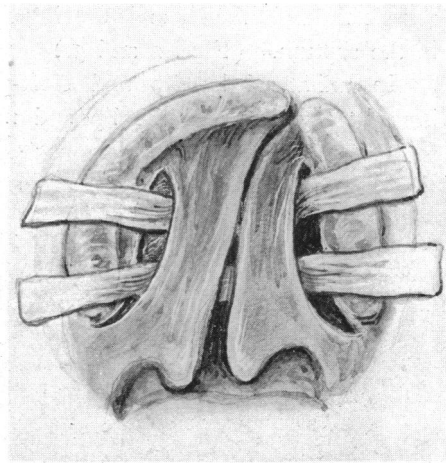

FIG. 26.—Semidiagrammatic sketch showing mucoperiosteal flap loosened on the patient's right side and a flap containing bone on the patient's left side. In the two-stage operation tapes are passed around both flaps and tied so as to hold the medial margins of the flaps in apposition, or nearly so, care being taken not to exert sufficient pressure by the tapes to shut off circulation. Iodoform gauze packs are used in the lateral incisions for twenty-four hours.

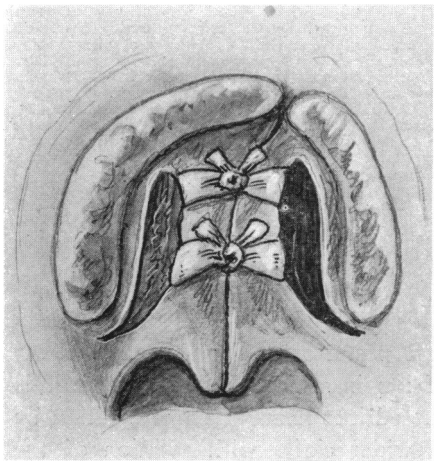

FIG. 27.—Semidiagrammatic sketch showing the tapes tied so as to hold the medial margins of the flaps in apposition. Iodoform gauze packs are used in the lateral incisions for twenty-four hours. One tape is removed on the third or fourth day, the remaining tape on the fifth or sixth day. On the seventh day the mucous membrane is removed from the margins of the clefts and sutures applied.

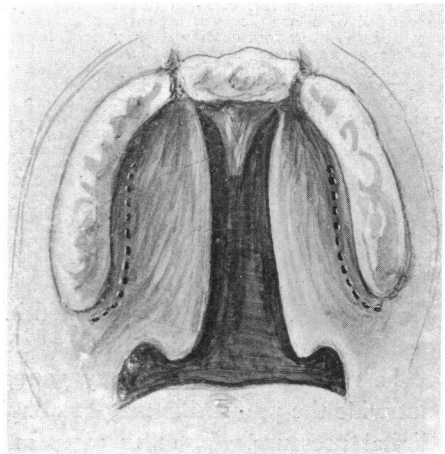

FIG. 28.—Semidiagrammatic sketch of bilateral cleft-palate, six or eight months after premaxilla has been placed in position and harelip repaired. Illustration shows the location of incisions which are carried down through periosteum, after which a thin narrow chisel is used to cut through the horizontal processes of the maxillæ and palate bones. Incision is then carried entirely through the mucous membrane forming the floor of the nostril, and the entire horizontal portion of the palate brought medialward.

Incomplete harelips must be made into complete ones by carrying the incisions into the floor of the nostril in order to get correct approximation of muscle tissue in the lip and to bring the ala of the nostril into proper relation to the septum. (Figs. 2 and 3.)

In unilateral harelip and cleft-palate the alveolar process should be brought together and the lip and nostril repaired preferably between the

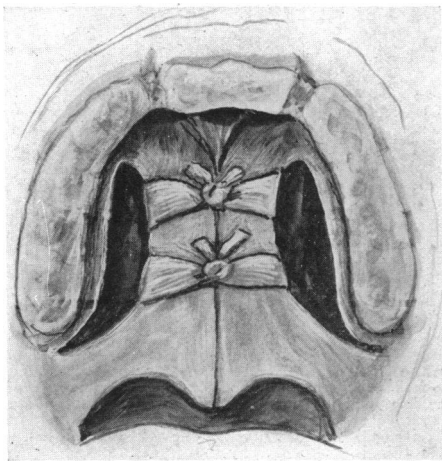

FIG. 29.—Semidiagrammatic sketch showing the method of loosening the flaps and bringing over bone in the repair of the double cleft-palate. The flaps are surrounded with two pieces of tape which are carefully tied so as to avoid cutting off any circulation. Iodoform gauze packing is used in the lateral spaces for twenty-four hours, both for the purpose of controlling any oozing and also to help hold the flaps in the desired position. One tape is removed on the third or fourth day, the remaining tape on the fifth or sixth day. On the seventh day the mucous membrane is removed from the margins of the clefts and sutures applied.

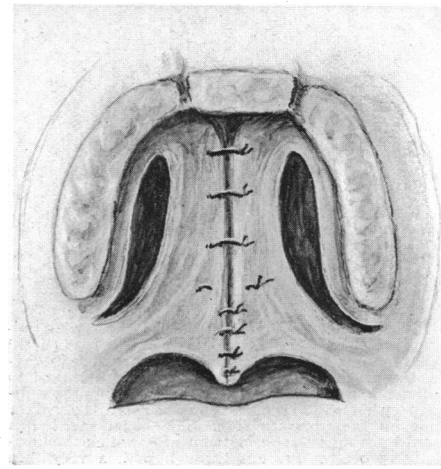

FIG. 30.—Semidiagrammatic sketch showing the application of sutures in a double cleft-palate in a two-stage operation. Double-ought wire sutures are used anteriorly, the sutures being carried through the entire thickness of the lateral flaps including the bone. These wire sutures are placed through the flaps before the mucous membrane is removed from the medial margins. This is important in that one is much less liable to detach the small strip of bone from its muco-periosteal covering. One on-end mattress suture is used near the junction of the hard and soft palate. Posterior sutures are black silk. Tapes are again placed around the flaps as shown in figure 29, using care in seeing that they do not produce much pressure on the flaps. One tape is removed usually on the second or third day, the remaining tape on the fifth or sixth day. Remove only one or two sutures each day beginning about the ninth day and having all sutures removed on the fifteenth day after operation.

second week and the fourth month. This part of the repair can usually be done at one operation, leaving the cleft posterior to the alveolar process to be repaired later. In very young infants in which the alveolar cleft is not wide the premaxilla may be simply forced into nearly normal position by digital pressure. In older cases and in wider clefts with rotation of the premaxilla, closure of the alveolar cleft is facilitated by partially dividing the outer portion of the alveolar process lateral to the canine area with a thin chisel, allowing a greenstick fracture of the inner portion of the process when the premaxilla is forced into position. A wire suture through the alveolar process holds the margins in apposition. (Fig. 4.) The lip is then repaired as in simple harelip. (Figs. 5 to 9.)

In bilateral harelip the philtrum should be preserved, bringing its inferior edge down to form the centre of the lip margin. (Figs. 12, 14, 18, 20, 24.)

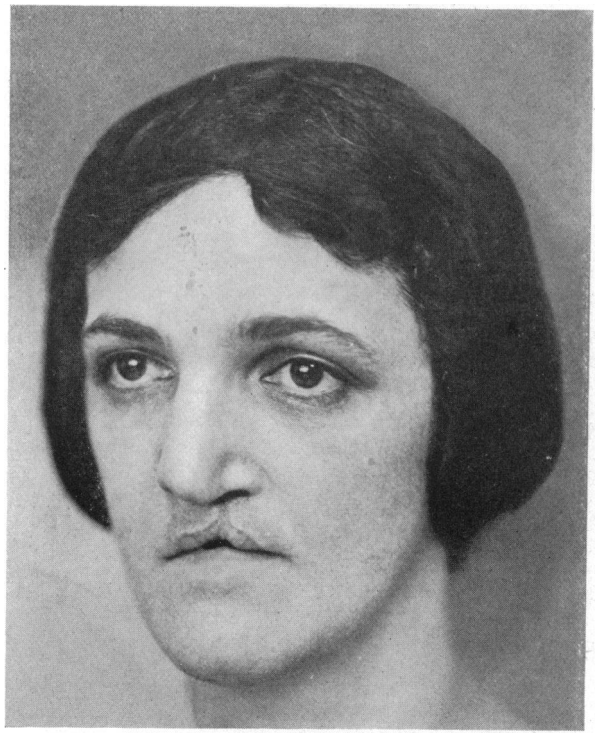

FIG. 31.—Case VIII. Age twenty-eight years. Unilateral harelip, bilateral cleft-palate. Lip had been operated upon when patient was two years old and again when sixteen years old. Note that vermilion border was not properly removed from margins of cleft in lip, persisting as a disfiguring red line extending to floor of nostril.

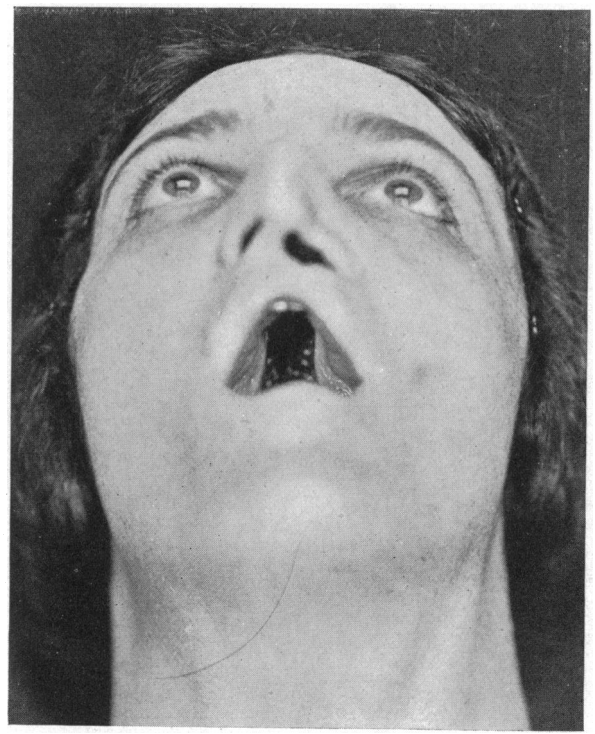

FIG. 32.—Case VIII. Showing deviation of the nasal septum, flattening of ala nasi and contour of nostril. Wide bilateral cleft of palate.

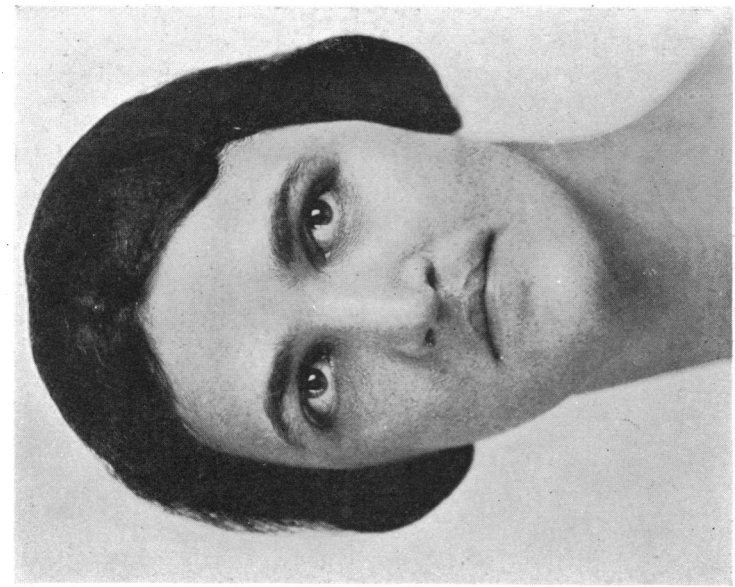

FIG. 34.—Case VIII. Showing contour of lip and nostril five months after second operation.

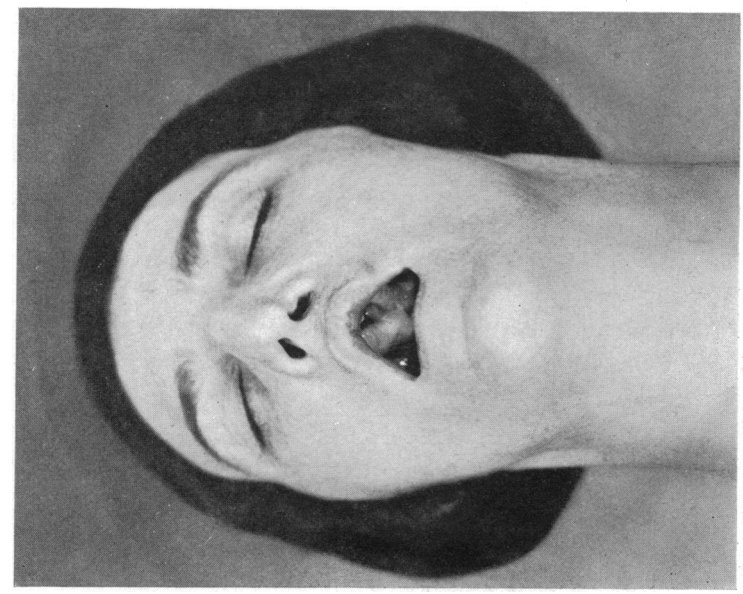

FIG. 33.—Case VIII. Showing condition seven weeks after operation on palate, four weeks after operation on lip.

Even when the philtrum is quite small its development after this method of repair is most surprising and gives a much better appearing and functioning lip than those repaired by trimming the philtrum to a V shape and approximating the lateral lip margins beneath it. We used the latter method for several years and recommended its use in several of our earlier papers, but the contracted appearance of the resulting lips in many of the cases caused us to discontinue its use.

In bilateral harelip and cleft-palate the vomer is elongated and the premaxilla shows varying degrees of superior rotation with corresponding shortening of the columella. When the deformity is marked the premaxilla can be placed in approximately normal position either by resecting a V-shaped section from the under surface of the vomer to allow postero-inferior rotation of the premaxilla (Figs. 18 and 19), or in less marked deformities the vomer can be split antero-posteriorly on the inferior surface of the anterior portion, allowing a bilateral bulging of the sides when the premaxilla is forced downward and backward into position where it is held by a wire suture.

After reconstruction of the alveolar cleft—unilateral or bilateral—and repair of the harelip, a period of several months is allowed to elapse before operating upon the remaining cleft. Such remaining clefts, and also palates with clefts not extending through the alveolar process, are preferably corrected between the ninth and the twentieth month. During the past eighteen months we have done all palate cases in which the clefts extended as far anteriorly as the midportion of the hard palate in two stages—the operations being done from five to eight days apart. In the majority of these cases we have brought the rudimentary horizontal processes of the maxillæ and palate bones over with the soft tissues, using the bone from the side not attached to the nasal septum in unilateral cases, and from both sides in bilateral cases. Operations bringing bone over with the flaps were devised originally by Ferguson, used in Philadelphia for many years by J. Ewing Mears and later revived, improved and more successfully used by W. J. Roe.

Bringing the bone over with the flaps is probably a more difficult operative procedure than to use the muco-periosteal flaps only. The ultimate results, however, we believe are distinctly better in those cases having bone in the flaps, the palates being better formed, maintain greater length, and have better function. One fact which has kept the operation from being popular has been the too frequent detachment of the bone from the mucoperiosteal flaps at the time of operation or else loosened to such an extent that it was subsequently lost. In such an event, if there is also failure of union of the mucoperiosteal portions, the necessary secondary operation is unquestionably more difficult and has less chance of being completely successful, than had mucoperiosteal flaps alone been used in the primary operation. However with our present technic of cutting and approximating the

flaps and routinely doing the operation in two stages, about one week apart, the bone is very seldom lost. The interval between operations allows time for the establishment of collateral circulation in the flaps which insures more certain and better union. (Figs. 25 to 30.) We prefer to have all the stages of harelip and cleft-palate operations completed before the child is two years old, since the cosmetic results and the quality of the speaking voice are best when the operations are done during that period. Gratifying results, however, are often obtained in older children or even in adults (Figs. 31 to 34), but the older cases are very apt to require longer training for the correction of the speech defect.