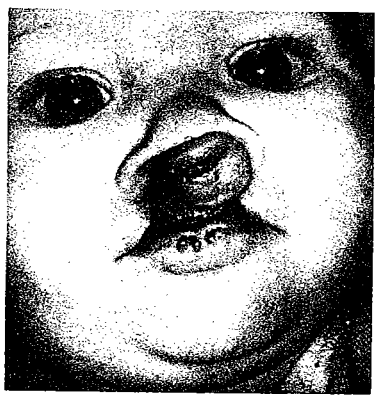
62. *Congenital Sinuses*

LOWER LIP

CONGENITAL lip sinuses, equally well known as mucous pits, were first described in the lower lip by DeMarquay in 1845. In 1951 Watanabe could find only 100 cases reported in the literature, but by 1967 Coccia and Bixler had found 200 and the numbers are mounting, as evidenced by Hoffman's 13 cases in 1971 from Mt. Sinai Hospital Cleft Palate Clinic, New York. Of course, many cases are not specifically reported. For instance, Viale-Gonzales, Barreto and Ortiz-Monasterio have so many clefts they can afford to include a case with mucous sinuses in association with a bilateral cleft presentation and not bother to report it separately.



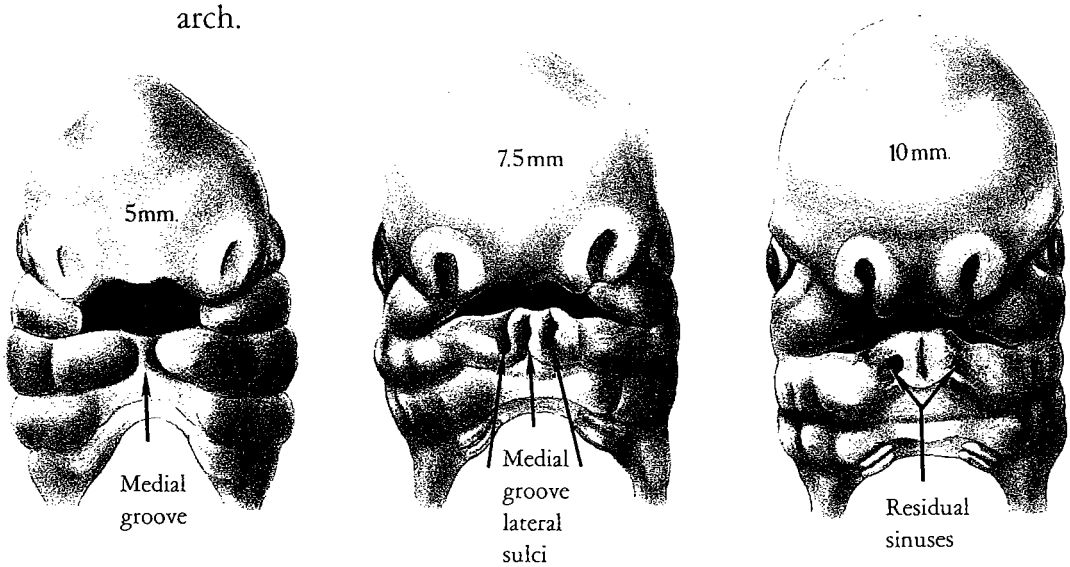
INCIDENTS OF ASSOCIATION WITH CLEFTS

In 1964 Gorlin and Pindborg found this anomaly associated with cleft lip or palate in approximately 70 percent, but a review of the world literature reveals that most authors find the association closer to 80 percent. In 1954 Van der Woude estimated that 0.5 percent of patients with cleft lip and palate also have mucous pits while Rintala, Lahti and Gylling estimated the association at 0.9 percent.

ETIOLOGY

Theories of the etiology of these sinuses range from intriguing to ridiculous. Sir Arthur Keith suggested that the phylogenetic origin might be found in the mucous canals of the lower lip of sharks, but Ludy and Shirozy failed to find any sharks with mucous canals. Other theories lay the blame on an attempt by the lower lip to close a cleft of the upper lip, amniotic adhesions, abnormal invagination of lip mucosa, faulty union of the mandibular processes and presence of epithelial pearls. Wang and Macomber considered each in detail and then struck them all off.

Evidently Steida in 1906 was the first to suspect the "sulci laterales labii inferioris." In 1912 Huber noticed in the human embryo a secondary notch on either side of the median groove of the lower lip after complete fusion of its two halves. In 1934 Sicher and Pohl found the presence of lateral sulci in 6.5 mm. embryos and, more clearly defined, in 9.2 mm. embryos; in later embryos these had disappeared. In 1952 Warbrick, McIntyre and Ferguson studied serial sections of human embryos and proposed that these sinuses were due to failure of obliteration of the cephalic end of the lateral sulcus of the developing mandibular arch.



Wang and Macomber in 1956 and S. Hoffman in 1971 favored this lateral sulcus theory. These congenital sinuses most often occur as a pair of dimples on each side of the midline. Yet they can occur unilaterally, usually on the left but occasionally on the right and very rarely in the midline, which condition seems

consistent with the theory of the persistence of the lateral sulci and, when in the middle, the median groove.

HEREDITY

Wang and Macomber considered the anomaly due to a single defective gene. Van der Woude did also, and in her series of five families, including 94 persons, these interesting findings appeared which are of value for genetic counseling:

1. Autosomal dominant inheritance with a 50 percent chance of inheriting the gene was shown.
2. The affected individual could have pits alone, cleft lip or palate alone, all three or any combination (and a few had no abnormality but could transmit the gene).
3. Of her 94 persons, 55 had abnormalities (pits and/or cleft lips and/or cleft palates) and 30 of the 55 had cleft lips and/or palate.
4. Persons most severely affected (i.e., having pits *and* cleft lips *and* cleft palates) transmitted more severe defects to their offspring: 25 percent had cleft lips and palate, 8 percent had cleft lip, 8 percent had cleft palate.

Yet as cleft lip and palate and cleft palate alone are believed to be different entities by Fogh-Andersen and congenital lip sinuses have been found in both these conditions, it may be more likely that two genes are involved. Test and Falls found lip sinuses in five generations of the same family. Others have reported families with various combinations of sinuses and cleft lip and/or palate which can be traced back for several generations. In 1943 Straith and Patton from Detroit reported a family of 13 persons which, over a span of three generations, produced six persons (or 50 percent) with bilateral cleft lip and/or palate. Each member with a cleft also had bilateral mucous pits of the lower lip secreting tenacious mucus from each of its pair of ducts opening just above the mucocutaneous line.

30-40% of children born into a family with lower lip pits will have CL P - 80-90% will have lower lip pits -

These anomalies, as noted, can be transmitted to approximately one-half of the offspring. Even an unaffected member of an involved family may have severely affected offspring. Cases

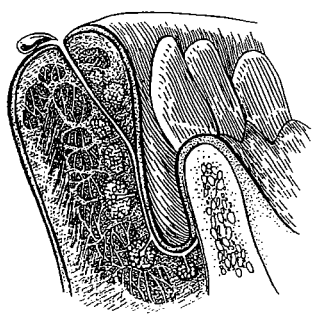
Robert Gorlin

reported in the literature suggest simple dominant inheritance with variable expressivity. In 1943 Fogh-Andersen first pointed out that the inheritance of clefts in the families with congenital lip sinuses is of a different character from that in families where no pits occur.

Robert J. Gorlin, Chairman of the Oral Pathology Division of the University of Minnesota, who also has an M.S. degree in chemistry, with J. Cervenka and S. Pruzansky, compiled a learned treatise for *Birth Defects* in 1971 entitled "Facial Clefting and Its Syndromes." They noted, in reference to lower lip pits and cleft lip and palate:

> The syndrome is transmitted as an autosomal dominant trait with 80% penetrance of any component of the syndrome but there is a possibility that the type of cleft present is influenced by modifying genes. The syndrome is seen with a frequency of about 1:75,000 to 1:100,000 live births and affects both sexes equally. On the basis of an analysis of 39 pedigrees, Cervenka, et al showed that an affected individual has a 22 to 39% chance of having an affected child with a cleft with or without lip pits. . . . Congenital lip pits of the same type have also been seen in association with the orofaciodigital syndrome and with the syndrome of popliteal pterygia.

A MUCOUS VESUVIUS



These dimples, which may appear as a circular depression or a transverse slit, are often situated at the apex of a nipple-like elevation. Each dimple is the orifice of a blind sinus extending downward and backward to penetrate the orbicularis oris muscle. The pit can vary in diameter from pin caliber to 2 mm., and its tract may extend in depth from 5 mm. to 2.5 cm. Lined by keratinized squamous epithelium like the vermilion with numerous mucous glands in the depth of the blind end, the pit may secrete a copious amount of mucus requiring wiping. Sir Arbuthnot Lane reported a case in which the secretion increased at mealtime.

MEDIAN LOWER LIP SINUSES

Although these sinuses most commonly occur bilaterally and anteriorly near the mucocutaneous junction, four median lower lip sinuses have been reported—by Ruppe and Magdelaine in

1927, Sato in 1938, Wang and Macomber in 1956 and Rintala and Lahti in 1973. Then there is a case reported by Rintala, Lahti and Gylling in 1970 and presented again in 1973 which was unusual in that the sinus was midline, relatively large and opening more posteriorly on the mucosal side of the lip. This sinus was found to be bipartite in its deeper part, with its septum covered by intact stratified epithelium. There was an associated bilateral cleft of the lip and palate. Oberst in 1910 reported a similar case. Miller in 1896 and Rose in 1868 also reported somewhat similar cases, in which the middle part of the lower lip resembled a nose with two nostrils from which the fistulous canals ran 2 cm. deep, each terminating in a separate cul-de-sac close to the mucosa of the labiogingival sulcus.

The occurrence of lower lip sinuses is often associated with minor facial developmental anomalies such as a short frenulum (found by Holbrook), sinus of the frenulum and fistula of the nasal bridge (MacKenzie) and preauricular tragal hillocks and occular dermolipoma (Parisien and Berken).

TREATMENT

In 1868 E. Rose suggested the indirect approach of an intraoral window to open the fistula into the oral cavity. In 1939 Baxter advocated more direct electrocoagulation of the entire tract. Mark Wang and Brandon Macomber, from experience with 15 cases, noted that the modern approach was complete surgical excision of the mucosal tract together with the surrounding glandular tissue. They warned that incomplete excision of the mucous glands would result in a mucoid cyst which in turn requires excision. They also advocated careful closure of the defect with special attention to the muscles to avoid undue looseness of the lower lip following orbicularis oris stretching from the presence of the sinus. Hoffman mentioned vertical elliptical excisions of the pair of sinuses in one of his cases.

Bill Lindsay in Mustardé's *Plastic Surgery in Infancy and Childhood* warned about mandibular lip pits:

The pits may be single or double, superficial dimples or deep sinuses. The

treatment is transverse elliptical excision including sinus opening and tracts. All the involved mucous membrane must be excised or the sinus will recur.

T. D. Rees and D. Wood-Smith in their elegant red and gold 1973 book, *Cosmetic Facial Surgery,* presented a case of upper and lower lip vermilion hypertrophy which also had a small midline pit surrounded by a tubercle in the vermilion of the lower lip. Paring of the excess vermilion and excision of the pit improved appearance but still enabled the patient to achieve a normal lip seal.

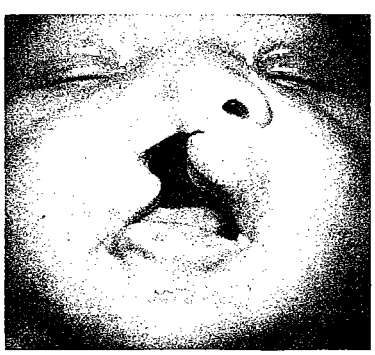
A. Rintala

The most fascinating description of lip sinus surgery was reported by A. Rintala and A. Lahti of the Finnish Red Cross Hospital in Helsinki in 1973 in the *Scandinavian Journal of Plastic and Reconstructive Surgery,*

After making an elliptical incision parallel to the lip around the lower lip sinus and elevating the edges of the sinus in order to facilitate dissection, it appeared that the sinus, extensive in its upper part, was divided into two at a depth of about 7–10 mm. Division was caused by a septum in the mid-line, like a pair of trousers. . . . Each "leg" continued separately almost to the bottom of the labiogingival sulcus for a distance of over 1.5 cm. The "legs" ran relatively close to the oral mucosa and converged toward the fundus, terminating close to one another in separate blind sacs from the bottom of which a fibrotic strand passed on to the anterior surface of the mandible.

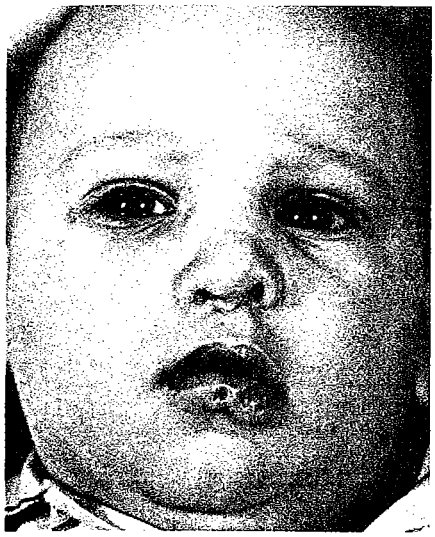
PERSONAL CASES

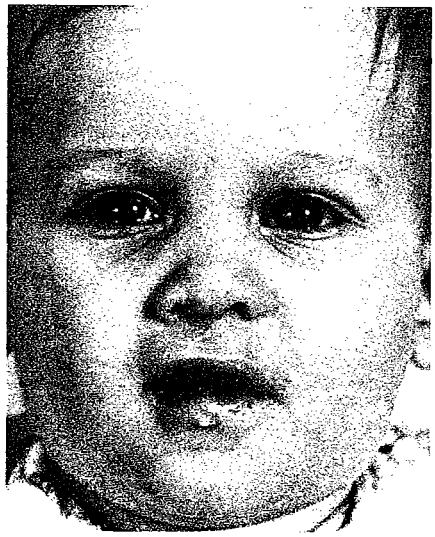
I have had several congenital mucous sinuses of the lower lip in cleft lip and palate cases. There has been nothing of special importance to note except that complete excision is not always easy. Of those I have treated, four are of varied interest. In two I treated the clefts and the sinuses primarily, in another the clefts primarily but the sinuses secondarily and in the third both the clefts and the sinuses secondarily.



One case of bilateral lower lip sinuses, first seen in 1963, was associated with a severe complete right unilateral cleft of the lip and palate. There was no family history of clefts or pits, but it can be predicted that there will be. The mucous sinuses were in the vermilion of the lower lip near the mucocutaneous junction on

either side of the midline. They were excised individually and transversely and the scars revised several years later.

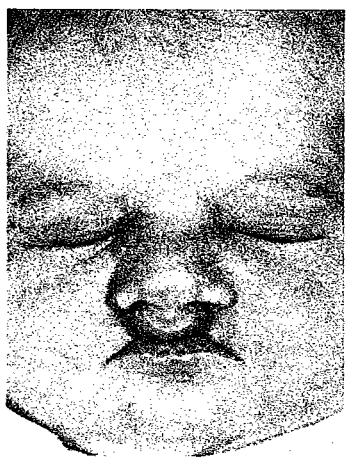

After R-A

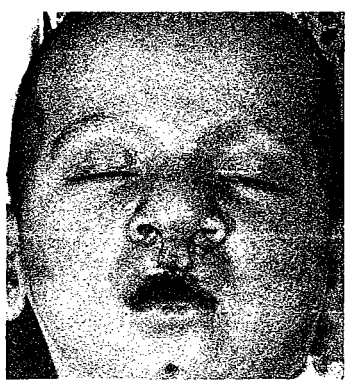

First excision of pits


Final excision


2 years later


Age 6 years


3 days



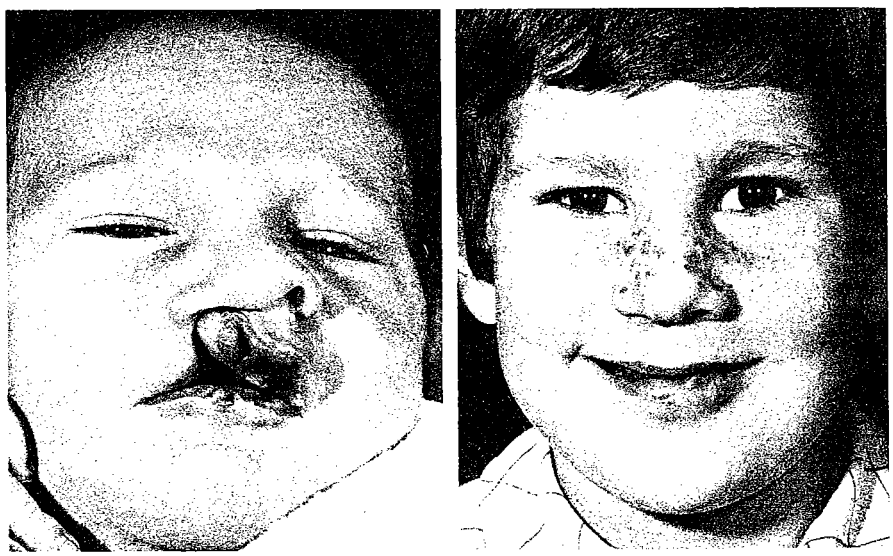
Another case of bilateral lower lip sinuses was seen in a girl with associated complete bilateral cleft of the lip and alveolus with protruding premaxilla but no cleft of the hard or soft palate. There was no history of clefts or pits in the family.

At 2½ months the premaxilla was set back and fixed in the notch in the hard palate after subperiosteal resection of a portion of the vomer. Lateral mucosa and muscles were joined behind the prolabium and a forked flap was banked as "praying hands" with the alar bases.

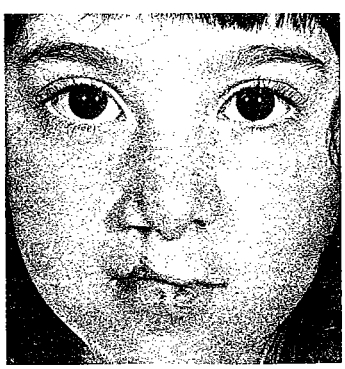
1 year

Eight months later, methylene blue was painted into the depths of the two pits and a transverse eliptical excision was extended 2 cm. deep into the lower lip dissecting out the sinuses like a pair of closed end trouser legs. The methylene blue facilitated total excision. The wound was closed in layers.

A boy was born in 1970 with a right complete cleft of the lip and alveolus associated with mucous sinuses of the lower lip. The mother had had a cleft of the lip and had a history of others in her family with clefts, but no mucous sinuses were reported. Rotation-advancement closure of lip was carried out at four months of age but sinus excisions were postponed. An attempt elsewhere resulted in right transverse and left oblique scars but persistent secretion of mucus.

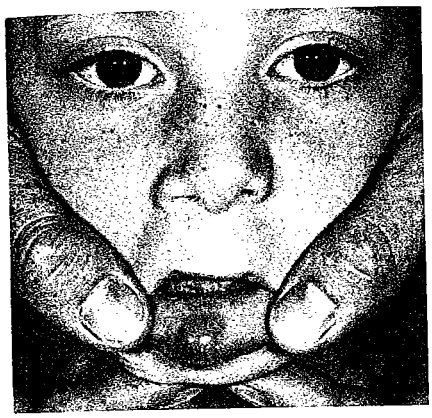


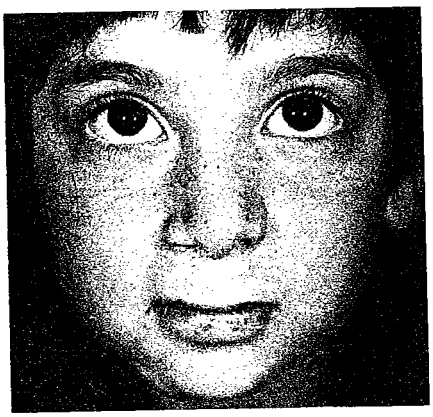
At age four years both scars were included in one large transverse excision carried deep to remove all mucosa of the sinuses. Wounds healed without difficulty, but the patient returned to South Africa. Although the father reported that all is well, no photographs have been forwarded as yet.



A baby girl was born in 1968 with right complete cleft of lip and alveolus with bilateral mucous sinuses of the lower lip. Lip closure and sinus excisions were carried out elsewhere. It is of particular interest that the great-grandfather, the grandfather and the father all have had mucous sinuses of the lower lip. It is also of importance that the patient has a normal fraternal twin.

In 1972 the lip and nose were revised and the mucous sinuses reexcised. A large mucous cyst formed postoperatively which required another, more extensive transverse excision resulting, finally, in complete removal of the intact mucocele and excellent healing.





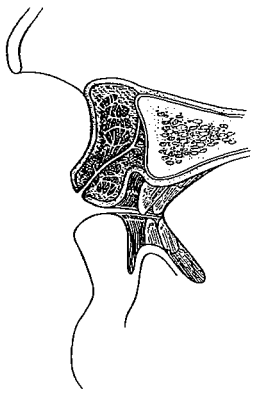
Total excision, of course, is the answer. If the sinus appears as a transverse slit, it may be easier to excise it horizontally, particularly if there is a pair. Occasionally better scar and contour may be achieved by excision of a vertical ellipse. Fortunately this anomaly, if handled correctly, leaves little to no residual deformity, and the importance of this outcome is multiplied as the defect has a 50-50 chance of reappearing in future generations.

MIDLINE SINUS OF THE UPPER LIP

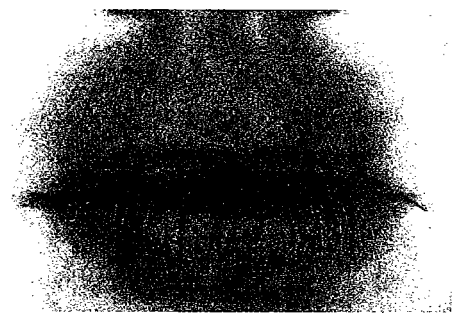
The earliest cases were reported by the French, Lannelongue and Menard in 1891 followed by Clavet in 1899. There are eight cases in the world literature; seven of the sinuses opened in the center of the philtrum and one at the base of the frenulum. MacKenzie's case had a 1 cm. blind tract not piercing the orbicularis oris ending near the nasal spine. Holbrook's case had a 2 cm. wide opening in the center of the philtrum surrounded by red epithelium. Microscopically these tracts have been seen to be lined with squamous epithelium. Clavet's case also had areas of columnar and polyhedral epithelium. In Holbrook's case the tract was surrounded by hyaline cartilage and, unlike the other cases, had

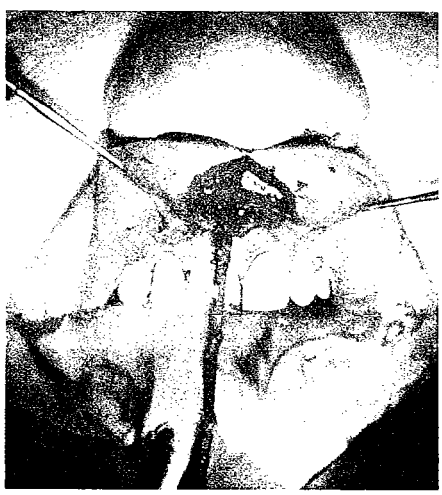
no hair follicles or sebaceous glands. Most cases had occasional mucus drainage, and two had a history of infection. There have been associated anomalies reported: Kriens' case had a double frenulum; MacKenzie's case had a cyst and fistula on the dorsum of the nose. Treatment, when recorded, has been excision of the entire tract.

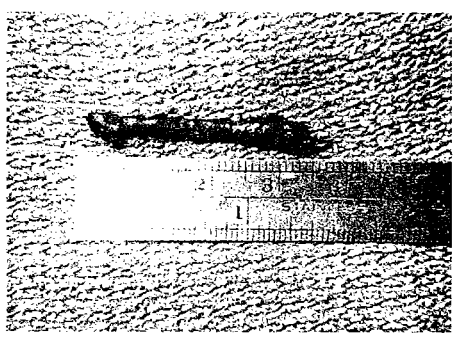
A VERMILION SINUS



Roger Bartels and Robert Howard of Orlando in December 1973 reported in *Plastic and Reconstructive Surgery* a case with mild hypertelorism and a minimal midline cleft of the vermilion of the upper lip with a tiny dimple at its center, the squamous epithelialized tract extending superiorly through the orbicularis oris muscle for 1 cm., then submucously for 2.4 cm., ending in a blind sac attached by a small fibrous band to the nasal spine. Total excision of the tract was carried out and followed by an uneventful recovery.







Bartels and Howard summarized upper lip sinus embryology in 1973:

Whether a midline sinus of the upper lip is a variation of (1) failure of downgrowth of the nasofrontal process, (2) breakdown of the mesenchyme-poor, fused, maxillary processes, or (3) failure of complete fusion of the maxillary processes growing together over the nasofrontal process, is not known. The association of hypertelorism with a midline sinus and cleft of the upper lip in our case supports the theories of Boyd and Frazer, that this anomaly is related to an abnormality of the nasofrontal process.

A DIMPLE IN THE DIMPLE

In 1960 I reported the incidental finding of a midline dimple in the skin of the medial element of the upper lip just above the mucocutaneous junction in a right complete unilateral cleft of the lip and palate. The tract extended several millimeters. It was observed as the medial component was rotated down into balanced position and then later simply excised.