Craniofacial malformations are relatively rare, and because of their multiple variations, it is difficult to classify them. The terminology has always been far from satisfactory, and there has been absolutely no unanimity in a system of classification up to now. Most authors have restricted themselves to an analysis of a more or less sharply defined area of the face. Tessier was one of the first who dared to break with the tradition in an attempt to establish a logical and orderly system for all the established craniofacial malformations. He considers a cleft to be at the basis of each of the malformations described. He argues that these clefts are situated along very definite axes, and he also stresses that although bone and soft tissue are rarely involved to the same extent, "a fissure of the soft tissues corresponds with a cleft of the bony structures, while the converse is also true."

In order to simplify the nomenclature of the clefts described, Tessier devised a system in which a number is assigned to the site of each malformation depending on its relationship to the sagittal midline. This system has become widely accepted in a very short time, because the recording of malformations is made easier and communication between observers is facilitated.

The system is, however, a descriptive, clinical classification that is not related to the embryology of the malformation, and understanding of the underlying pathology is not really increased. In fact, the use of the word clefting may even create confusion where no cleft exists. For example, De Myer considers a cleft to be "a defect in apposition of structures along a junction." This concept is easy to accept when it refers to a primary or transformation defect caused by the failure of fusion of junctional structures, but it becomes unreliable when it refers to secondary or differentiation defects, such as oro-ocular clefts (Morgan II and III), which cannot be explained by classical embryologic theories, and it can certainly not be applied to such malformations as teleorbitism, Treacher Collins syndrome, hemifacial microsomia, and so forth. These malformations may in fact be associated with a cleft, but basically none of the cleft characteristics are present. Therefore, we feel that the word cleft is a misnomer because it makes it impossible to include such malformations as nasal aplasia, with or without proboscis, anophthalmia, or microtia. As a result, the Tessier classification becomes incomplete.

The objections mentioned here can be overcome by the embryologic classification we are about to propose and in which we have tried to emphasize a correlation between clinical observation and morphogenesis. This classification provides a framework which, on the one hand, is based on solid knowledge—gained either from the literature or from personal experience—and, on the other hand, leaves enough flexibility to be modified by future additional information.

**IDENTIFICATION OF CRANIOFACIAL MALFORMATIONS**

As a general rule, no classification is possible without having clearly identified the individual phenomena to be classified. This requires, first of all, an agreement on the terminology. As a second step, an accurate study of the similarities and dissimilarities is essential. Deductions can be drawn afterwards.

Some of the craniofacial malformations are identified by the name of the author who first described them (Goldenhar, Pierre Robin, and so forth). Such a name may convey a meaning to
the insider but is easily forgotten by the layperson. Other malformations are identified by their appearance and have been given such names as a cleft, hemifacial microsomia, retromandibulism, hypertelorism, and so forth, thus risking to create the impression that these malformations have a different etiology, which, in fact, may not be true.

The ultimate form of the craniofacial skeleton is determined by the growth or lack of growth in such functional areas as the cerebrum, the eye, the nose, and a number of ossification centers. When one wishes to include all craniofacial malformations in a classification and also use a common denominator, dysplasia is the only term that is applicable.

An arrest in skin, muscle, or bone development, no matter what the etiology, will manifest itself in "focal fetal dysplasia." The consequences of this dysplasia, or, in other words, its ultimate appearance and severity, therefore depend on the localization and the time of the disturbance.

Identification by the Localization of the Malformation

To allow for proper identification, the dysplasia should be named after the area or areas (facial processes or bones) involved. Modern knowledge of the normal embryonic and fetal growth processes has increased our understanding of the underlying pathology and provides the basis for a morphogenetic classification.

Identification by the Time of the Developmental Arrest

Whether or not the cerebrum and/or the eyes are involved has a most important bearing on the severity of a malformation. A developmental arrest of the forebrain will affect the development of the craniofacial skeleton and is incompatible with normal life. This aspect should receive attention in a classification, and distinction should therefore be made between groups with or without cerebral involvement.

Furthermore, the severity of a malformation will depend to a considerable degree on whether dysostosis or synostosis is present. The difference between the two should be stressed and translated into a second subdivision between craniofacial malformations with dysostosis and those with synostosis. In malformations characterized by dysostosis, distinction can again be made between transformation defects, which are caused by a developmental arrest occurring before or during the fusion of the facial processes (≤ 17 mm C.R.L.) and differentiation defects, which originate after this period.

The first category consists of skeletal malformations produced by focal dysplasia of growth centers such as the eye vesicle and nasal placode and/or by lack of cell degeneration between the facial processes resulting in primary clefts. The second category represents defects that are caused by defective differentiation and outgrowth of bone centers and cartilage, such as hemifacial microsomia and the secondary clefts. These differentiation defects may vary widely in their appearance, because they develop only when the ectoderm of the face has closed. The fact that the appearance of a secondary cleft is different from that of a malformation such as hemifacial microsomia can be easily understood when one visualizes what may happen when growth is arrested in one area while it continues in the normal surrounding tissues. An hour-glass deformity may develop, with the developmental arrest in the middle behaving as a scar that prevents the surrounding tissues from growing normally (Fig. 1).

Depending on the particular qualities of these tissues a series of defects may be produced such as colobomata of the lower eyelid and upper lip,

![Fig. 1. Morphogenesis of maxillary dysplasia (oro-ocular cleft production).](image-url)
an alar cleft, or an irregularity of the hairline (widow's peak). Finally, after comparing one craniofacial malformation with another, it has become obvious that all these malformations can be graded and identified with certain stages of embryonic life.

**Materials and Methods**

The present study is based on an intensive search of the literature on craniofacial malformations and on observations of a great number of patients seen at the Departments of Plastic and Reconstructive Surgery of the University Hospitals of Milan, Rotterdam, and Nancy, respectively. Diagrams of representative cases have been made, showing the subdivision of the classification based on our knowledge of the embryology.

**Classification of Craniofacial Malformations**

**Cerebral Craniofacial Dysplasias**

*Interophthalmic dysplasia.* We have grouped under this heading cases with either agenesis or hypodevelopment of the midline structures of the face and brain. They have in common an absence or severe hypoplasia of the premaxillary nasal and lacrimal bones, nasal septum, and the ethmoid with crista galli. The combination in different degrees of the preceding defects with hypotelorism is pathognomonic of a brain that has failed to divide into cerebral hemispheres (holoprosencephaly) and that normally lacks olfactory bulbs and tracts (arhinencephaly). The deformities belonging to this group have been widely reported in the literature. At one end of the range one finds cyclopia, synophthalmus, or synorbitism with holoprosencephaly and complete absence of the midline facial structures. From this stage, the spectrum of cerebral craniofacial anomalies with hypotelorism moves through ethmooencephaly, cebocephaly, and median cleft lip without a premaxilla toward the normal states.

*Ophthalmic dysplasia.* Malformations of the eye are quite common, such as anophthalmos, microphthalmias, and coloboma of the eyewall. They may be found in combination with many of the other dysplasias to be described.

**Craniofacial Dysplasias**

*Dysostoses.* Chronologically, development of the craniofacial skeleton proceeds along a helical course, symbolized by the letter S (Fig. 2). It starts with the formation of the middle and anterior cranial fossae in a posteroanterior direction and with the reduction of interorbital distance. It is followed by the growth of the nasomaxillary complex, expanding forward, downward, and laterally, and it is completed by the lengthening of the mandibular ramus that is produced by this expansion. Since the anterior projection of the greater wing of the sphenoid seems to play an important role in this development, we have chosen to start the craniofacial helix, in the lateral-posterior wall of the orbit, at the junction of the sphenoid and frontal bone. The upper half of the S encircles the orbit and the lower half of the S

![Cranio-Facial Dysplasias](image-url)
**TABLE I**

Periocular Dysplasias

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encircles the mouth. Dysplasias in the upper half of the S may be associated with ocular and periocular dysplasia (Table I). Dysplasias of the lower half of the S are frequently betrayed by the presence of preauricular tags, pits, and fistulas. Combinations of dysplasias in adjoining areas are described in this section, but only after due attention has been given to a description of the dysplasias separately. Combinations of dysplasias in separate areas of the face that are not in continuity with the S are not discussed.

Bilateral occurrence of similar or different malformations and monolateral occurrence of different malformations are all possible and in fact have been described. When encountered, however, they can now be identified with more ease.

**Sphenofrontal dysplasia.** Our review of the different forms of craniofacial dysostosis starts with malformations of the sphenofrontal area (Fig. 3). It is in this area that dysostosis and synostosis overlap with each other in a curious way and a clue to the delineation of the two pathologic processes may eventually be found. Comparison of rare cases of bony "cleft" formation (Tessier No. 9 cleft) in the sphenofrontal and sphenozygomatic area with the cranial grooves, which one can observe in "clover leaf" skull malformations (Fig. 3) and to a lesser degree in patients with plagiocephaly, brings us to the conclusion that there is a striking similarity in the localization of these malformations, stimulating speculation as to their cause. Unfortunately, too little is known about their pathology.

**Frontal dysplasia.** Orbital hypertelorism or teleorbitism and nasal dysplasia are frequently associated with defects in the frontal bone. A widow's peak and dystopia of the eyebrow may also be observed. Depending on the location and severity of the bony defects, a variety of encephaloceles may result (Fig. 4). In his classification, Tessier distinguishes between clefts No. 10 and 11, assigning to each a specific location in the frontal bone. We find it difficult to accept that the location and form of these defects are subject to a rule and believe that they are determined by the degree of growth retardation within one of...
the paired centers of the frontal bone. This means that they may be found anywhere, as an isolated frontal bone defect or in continuity with areas of deficient ossification in the nasal or maxillary bones.

Frontofrontal dysplasia. Bone defects with or without encephaloceles are sometimes found in the midline between or below the two halves of the frontal bone (Fig. 5). They may occur in combination with frontonasoeothmoidal and internasal dysplasias.

Frontonasoeothmoidal dysplasia. In the literature, the name frontonasal has been given to a wide variety of malformations occurring in the frontonasal process. We believe it to be a misnomer because it is not sufficiently specific, and we suggest that the term frontonasal or frontonasoeothmoidal should be reserved for developmental activities (including their absence) at the junction of the frontal and nasal or ethmoidal bones.

When, for example, reduction of the interorbital distance does not occur and a hypertelorism situation (Fig. 6) is retained, we feel justified in using the term frontonasoeothmoidal dysplasia. Whatever the cause of the orbital hypertelorism or teleorbitism, it is certainly not a cleft. Widening of the ethmoid bone, its cribriform plate, or its crista galli are not necessarily symptoms of a cleft. To us these features merely represent the effects of a deficient development of the nasal capsule, which may allow for further growth but not for further modeling by narrowing and/or elongation of the bony structures involved.

Internasal dysplasia. Internasal dysplasia (median cleft nose, bifid nose, doggenase, No. 0 cleft) represents a group of malformations in which two nasal halves of normal appearance are separated by a groove (Fig. 7) that may be wide and shallow or narrow and deep. This condition is frequently associated with a median cleft lip. Orbital hypertelorism or teleorbitism is always present in the more severe cases, its degree increasing with the width of the internasal defect or groove. The premaxillae are not absent, but they may be retarded in development and not fused (premax-
ilopremaxillary dysplasia). The maxilla may show a keel-shaped deformity with the incisors rotated upward in each half of the alveolar process.

Sometimes a medial cleft is also found in the palate, which has the form of an inverted V and may extend up to the cribriform plate. The distance between the palate and the cranial base is extremely short, and in the remaining internasal space, little else may be found but an undifferentiated cartilaginous mass. A widow’s peak is frequently observed.

**Nasal dysplasia.** Since embryologically the nose is made of two distinct nasal halves and the majority of the nasal malformations are restricted to one of these halves, it seems appropriate that the name nasal dysplasia should be used only to indicate a unilateral malformation, while the word rhinal should be applied when both halves are involved. Nasal malformations are extremely rare, and it is possible to distinguish between four different types:

1. Nasal aplasia
2. Nasal aplasia with proboscis
3. Nasoschizis
4. Nasal duplication

**Nasal aplasia.** This malformation is characterized by complete absence of one nasal half (Fig. 8). When both halves are absent, the term arhinia should be used. In nasal aplasia, there is no nasal cavity and the cribriform plate, the olfactory bulb, and the nasal bone are similarly missing. Pneumatization of the maxillary, ethmoidal, sphenoidal, and frontal sinuses has failed, and exploration reveals nothing but solid bone. There is no nasolacrimal duct; instead, cyst formations or infections in the deformed lacrimal system are frequently encountered. The affected half of the maxilla may be hypoplastic, and the palatal vault may be high and acutely arched. The premaxillae are relatively normal.

**Nasal aplasia with proboscis.** These are the malformations in which aplasia—with all its characteristics—is associated with a proboscis (Fig. 9). The origin of this club-shaped appendix is commonly found at the upper inner canthal region. Examination of the proboscis may reveal the same tissues as are found in a normal nose: hair, sebaceous glands, sweat glands, striated muscle fibers, nerve fibers, and cartilage. There is, however, one major difference: The proboscis does not contain a cavity but a narrow tract with a mucous lining that ends blindly at the level of the dura mater or at the cranial base, where the cribriform plate and olfactory tracts are missing. Tears have been known to issue from this tract, but their origin is not clear. They may have been produced by lacrimal gland tissue surrounding the tract or have come from a communication with a lacrimal apparatus.

**Nasoschizis.** Nasoschizis (Tessier’s No. 1 cleft) is characterized by a deformity of one-half of the nose in the presence of a normal septum and a nasal cavity (Fig. 10). This malformation is probably not as rare as one might expect from the scarce literature on this subject, since Mazzola
was able to add 14 cases of his own. The severity of the malformation ranges from a more or less complete cleft of one-half of the nose with absence of the nasal bone to minor deformities of the ala consisting of one simple notch or even two notches. Severe nasoschizis, teleorbitism, and a widow's peak are frequently found together, resembling the typical hour-glass deformity, which also may be produced by maxillary dysplasia.

**Nasal duplication.** Duplication of one-half of the nose (Fig. 11) is also known to occur in different forms. Mazzola's cases show that the scale of variations may range from a bifid nostril to complete rhinal duplication.

**Nasomaxillary dysplasia.** This category consists of two groups. The first consists of the lateral nasal maxillary dysplasias, which have their origin at the junction of the lateral nasal and maxillary processes. The second group consists of the medial nasal maxillary dysplasias, which are found at the junction of the medial nasal and maxillary and lateral nasal processes.

In lateral nasal maxillary dysplasias, distinction should be made between transformation defects that develop before fusion of the lateral nasal and maxillary processes has taken place and differentiation defects that are found after the ectoderm has closed. Transformation defects consist of a real ectodermal and bony cleft between the lateral nasal and maxillary processes along the nonfused nasolacrimal groove. In these cases, the nasolacrimal duct is absent or partially developed. Differentiation defects represent malformations that are caused by deficient ossification occurring after the fusion of the lateral nasal and maxillary processes. They are characterized by a shortening of the distance between the alar base, which is found in a cranial position, and the medial canthus, which is dislocated downward. The involvement of the maxilla can be deduced from such deformities as canthal dystopia, dysostosis of the frontal process of the maxilla, and caudal displacement of the orbital floor.

In medial nasal maxillary dysplasias, distinction can again be made between transformation and differentiation defects. A transformation defect is characterized by an ectodermal and bony cleft between the medial nasal and maxillary and lateral nasal processes. The cleft lip thus produced is frequently found in combination with other craniofacial dysplasias, but with lateral nasomaxillary dysplasia (Fig. 12) it forms the well-known malformation also known as Morian I, Tessier No. 3, or naso-ocular cleft. Gunter, however, coined the term “nasomacillary.” Differentiation defects are recognizable as a bony cleft between premaxilla and maxilla (premaxilla-maxillary dysplasia). The ectoderm of the lip is closed.

**Maxillary dysplasia.** The medial oro-ocular cleft (No. 4 cleft of Tessier or Morian II “cleft”) and the lateral oro-ocular cleft (No. 5 cleft of Tessier or Morian III “cleft”) are two of the commonly used terms for a malformation in that part of the craniofacial skeleton that has been formed by the maxilla.

The medial oro-ocular cleft (Fig. 13), called in this classification *medial maxillary dysplasia*, runs
from the medial third of the lower eyelid to the upper lip, midway between the philtral crest and the labial commissure. The nose is left free, and in extreme cases, the position of the nasal nostril aperture is at the same level as the eyeball. Contact between the edges of the cleft varies. Sometimes there is a huge gap with nothing to separate the contents of the orbit from the mouth. More commonly, however, there is an epithelial bridge connecting the colobomata of the lower eyelid and the upper lip. The skeletal malformation is generally considered to consist of a cleft. In the maxillary body, this cleft is situated medially to the foramen of the infraorbital nerve. In the alveolar process it proceeds between the lateral incisor and the canine tooth. However, the existence of a bony cleft cannot always be demonstrated.

In a patient of the first author, the malformation of the soft tissues was not associated with a bony cleft, but with severe hypoplasia of the maxilla. The same observations were made by Miller et al.

The lateral oro-ocular cleft (Fig. 14) connects mouth and orbit by colobomata in the lateral third of the lower eyelid and the lateral third of the upper lip. In the maxillary bone, the cleft runs laterally from the infraorbital foramen, justifying the term lateral maxillary dysplasia. These malformations are extremely rare, and consequently, there has been little opportunity to study their morphology.

Morian and Tessier observed that the bony defect on the orbital side was located at the body of the maxilla, while on the alveolar side it was situated between the canine tooth and the premolar. These findings suggest that the malformation is restricted to the maxilla, implying that we are dealing with a pure maxillary dysplasia. The similarity between the localization of the soft-tissue malformation found in lateral maxillary dysplasia and Treacher Collins syndrome may, however, be so striking that there is reason to suspect that at least in some of the so-called
lateral oro-ocular clefts, the zygoma is also involved.

Duhamel\textsuperscript{14} in fact has stated that the maxillary dysplasia in patients with a lateral oro-ocular cleft is frequently associated with a malformation of the malar bone. Duke-Elder\textsuperscript{15} even names these malformations “maxillozygomatic dysplasias.”

Maxillozygomatic dysplasia. This is the No. 6 cleft of Tessier,\textsuperscript{1} which he considers to form an integral part of the Treacher Collins syndrome. In the skeleton, a defect is found between the maxillary and zygomatic bones. In the soft tissues, the dysplasia is marked by the existence of a groove that runs from the lateral third of the lower eyelid downward in the direction of the corner of the mouth or more laterally.

The orientation of this groove is virtually identical to that of the soft-tissue malformations found in lateral maxillary dysplasia and in Treacher Collins syndrome, suggesting that the clinical appearance is not always sufficient to predict the nature of the underlying osseous malformation and that diagnosis is only possible after inspection of the skeleton. These patients are rare, data are scarce, and more information is urgently needed. For instance, it may well be that Pitanguy’s\textsuperscript{16} case with bilateral clefts should not be classified as a lateral maxillary dysplasia or a Tessier No. 5 cleft, but as an example of bilateral maxillozygomatic or even zygomatic dysplasia. An oral view of this patient gives the impression that the bony defect is not situated in the maxilla itself, but in the zygoma.

Hovey et al.\textsuperscript{17} correctly noted the resemblance between a patient of Rogers with Treacher Collins syndrome and Pitanguy’s case. In a patient of the first author with a Treacher Collins syndrome, the course of the deep grooves between the lateral corner of the mouth and the lateral part of the lower eyelid was similar to that of the grooves found in a No. 5 cleft. At exploration, however, the malformation proved to be caused by an absence of both malar bones.

Zygomatic dysplasia. Malar hypoplasia is the hallmark of the malformation known as Treacher Collins syndrome. The dysplasia of the malar bone is usually associated with antimongoloid angulation of the palpebral fissures, notching of the lateral part of the lower eyelids, partial absence of eyelashes in the lower eyelids, flattening of the cheeks, and occasionally, a groove running from the lateral part of the lower eyelid to the corner of the mouth or more laterally. When no other symptoms are found, it seems justified to speak of the incomplete form of the syndrome, which, with few exceptions, is found bilaterally (Fig. 15).

Zygofrontal dysplasia. This is the No. 8 cleft of Tessier,\textsuperscript{1} which can be observed in Treacher Collins and Goldenhar syndromes. The cleft is obviously the result of a developmental arrest in the fusion, i.e., suture formation, of frontal bone and zygoma. It may be characterized by the presence of an epibulbar dermoid and the absence of a lateral canthus.

Zygotemporal dysplasia. Clefts at the borderline between zygoma and temporal bone (No. 7 cleft of Tessier) can be found in Treacher Collins syndrome as well as in hemifacial microsomia. The malformation may be associated with a dysplasia of the temporal muscle and with anterior displacement of the sideburns.

Temporoaural dysplasia. The word aural signifies everything pertaining to the ear, i.e., the auris. The term temporoaural focuses attention on the intimate developmental relationship in time and space between parts of the temporal bone and the different aural structures.

Distinction can be made between dysplasias of the external ear, the middle ear, and the inner ear. The malformations that result can occur separately or in combination with each other. They are representative of the stage at which normal development was disturbed, and comparison of auricular malformations with the different stages of development makes it clear that major deformities, such as microtia, reflect an arrest or an early disturbance of the embryonic development of the external ear, while other deformities,
such as lop-ear and cup-ears, are clearly generated at a later stage.\textsuperscript{18} Temporoaural dysplasia may be associated with a paralysis of the facial nerve.\textsuperscript{18}

Zygotemporoauromandibular dysplasia. This is the complete form of Treacher Collins syndrome. It was described by Franceschetti, Zwahlen, and Klein,\textsuperscript{20,21} who referred to it as "mandibulofacial dysostosis." In this malformation, the characteristic features of zygomatic dysplasia are associated with those of (1) zygotemporal dysplasia (forward displacement of sideburns and deficiencies of the temporal and masseteric muscles), (2) temporoaural dysplasia (malformations of the external and middle ear, rarely of the inner ear), and (3) mandibular dysplasia (malformation of the condylar and coronoid processes, deficiencies of the mandibular ramus, and obtuse angulation and antegonial notching\textsuperscript{22} of the mandible (Fig. 16).

Like the incomplete form, zygotemporoauromandibular dysplasia is almost always bilateral. The syndrome may be associated with malformations of the eye, the nose, and the mouth. Pruzansky,\textsuperscript{22} however, reported that the degree of external ear malformations does not correlate with hearing function. In most patients, the malformation is unilateral. The incidence of bilateral to unilateral cases ranges between 1 to 6\textsuperscript{23} and 1 to 18.\textsuperscript{24} The malformation may be associated with deficiencies of the temporal, pterygoid, and masseter muscles, with facial nerve anomalies and with hypoplasia of the parotid gland. Abnormalities of the palate, maxilla, zygoma, and vertebrae may also exist.

Temporoauromandibular dysplasia. Auromandibular dysostosis, hemifacial microsomia,\textsuperscript{25} branchial arch syndrome,\textsuperscript{26} and auriculobranchiogenic dysplasia\textsuperscript{27} are but a few of the terms used to indicate this syndrome, of which condylar anomalies seem to be the hallmark. A whole scale of variations (Fig. 17) is possible, ranging from temporoaural malformations of maximal severity, coexisting with mandibular deformities that are scarcely apparent, to minimal malformations of external and middle ear associated with characteristic maldevelopment of the mandibular ramus and condylar process.\textsuperscript{24}

A radiographic analysis of first and second branchial arch anomalies brought Caldarelli and Valvassori\textsuperscript{28} to the conclusion that the severity of external ear malformations parallels the severity of mandibular malformations, although not necessarily in terms of gradation specificity. According to these authors, there is also a parallel between the severity of ossicular and auditory canal dysplasia with the severity of either the external ear or mandibular malformation.

Maxillomandibular dysplasia. A failure of the maxillary and mandibular processes to fuse results in macrostomia, a malformation characterized by a transverse "cleft" running from the angle of the enlarged mouth to the tragal area (Fig. 18). Most of these clefts are associated with preauricular appendages or fistulas, but other temporoaural dysplasias (middle ear deformities, etc.) and maldevelopment of the mandible may
also occur. Unilateral as well as bilateral cases have been described.

Mandibular dysplasias. Best known in this group is the Pierre Robin anomaly (Fig. 19), which consists of micrognathia, glossoptosis resulting in respiratory distress, and cleft palate. Two types of micrognathia exist. In the first type, the mandible is small and remains small. In the second, rapid growth is seen after birth, resulting in a normal mandible. Micrognathia should, however, be distinguished from retrognathia, in which a mandible of normal size is held back by abnormal musculature. For an excellent review of this malformation, the reader should consult the work of Randall and Hamilton.

Intermandibular dysplasia. This malformation completes the list of craniofacial dysplasias. Clefts of the lower lip and mandible are clearly caused by a failure of the two mandibular bone centers to fuse, i.e., a secondary defect (Fig. 20).

Synostoses. Craniofacial malformations with synostosis can be purely cranial or facial or they can be of a craniofacial nature. Craniosynostosis comprises a group of malformations that are usually classified by their appearance and known under the following names: dolichocephaly, brachycephaly, acrocephaly, triogonocephaly, and plagiocephaly. To many, these words convey no meaning, and it would therefore be wiser to use names that refer to the localization of the affected sutures, such as sagittal dysplasia, coronal dysplasia, sagittocoronal dysplasia, metopic or frontal dysplasia, and monolateral coronal dysplasia, thus following the advice of Bertelsen.

Whether synostosis is a secondary manifestation of an underlying malformation in the basi-cranium, as Moss and Stewart et al. want us to believe, or a primary event in the sutural tissues, as has been postulated by Parks and Powers and recently by Cohen, is still not
known. We tend to agree with Cohen, that whatever mechanism responsible for progressive calcification throughout the body—as in Apert’s syndrome—should also be held responsible for premature craniosynostosis. As one of us said, “The sutural tissues are too old too early.”

However, the relation between dysostosis and synostosis and the role played by each of these processes in the production of a malformation will have to be specified before accurate classification of craniofacial malformations with synostosis becomes possible. This applies especially to facial malformations, since premature fusion of bone centers in the face is even more difficult to diagnose.

**DISCUSSION**

Classifications of common and rare craniofacial clefts are almost always based on their localization, i.e., their topography.1,7,37–39 Some authors divide the face into more or less vertical regions,7,38,39 others group the defects around the brain, sense organs, and the branchial arch system,40 and still others1,3,41,42 confine themselves to a description of some rare defects of the face. Tessier1 introduced a cleft numbering system of practical, clinical value from 1 to 14 in which the orbit takes a key position. None of these authors classified on an embryologic basis, because some clefts could not be explained in terms of development.

To facilitate communication and avoid confusion the word *dysplasia* is advocated as the common denominator for all the malformations to be discussed. The localization of each malformation is indicated with the name of the developmental area or areas (facial processes and bones) involved. From the embryologic point of view, a distinction must first of all be made between cerebral craniofacial defects involving the brain and/or the eyes and defects of the face and cranium, the so-called craniofacial defects. The cerebral craniofacial malformations develop very early in embryonic life.5 The different types of the holoprosencephalon group5 have in common absence of the area between the nasal placodes. The anlage of the two eye vesicles occurs in separate eye fields, and they fuse in the facial midline during and after the closure of the neural tube in that area. This developmental view is contrary to the general idea that the cyclopic condition is the result of the faulty organization of paired optic centers in a common eye field.42

The subdivision of craniofacial malformations into dysplasias with dysostosis and dysplasias with synostosis is indicated because the localization of areas with premature synostosis corresponds with the sutural defects of dysostosis. The pathomorphogenesis of synostosis is now under study.

Craniofacial malformations with dysostosis may originate before the facial processes have fused and ossification has started (±17 mm C.R.L.). They are then called *transformation defects*. Nasal aplasia and the primary clefts that continue to separate the facial processes belong to this category. However, dysostosis may also be produced by deficient ossification of bone centers after the primitive face has formed. The malformations then produced are named *differentiation defects*. Theoretically, these defects can be caused by the absence of the “anlage” of bone centers or by insufficient outgrowth of these centers.2

**SUMMARY**

A new classification of malformations of the face and cranium is proposed, based on embryologic studies and observations concerning a great number of patients seen by the authors. First of all, one should distinguish between cerebral craniofacial (with brain and/or eyes involved) and craniofacial malformations. Craniofacial malformations may be characterized by dysostosis and by synostosis. Malformations with dysostosis may be produced by transformation as well as differentiation defects. Synostosis is always caused by a differentiation defect. A new nomenclature is introduced.

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