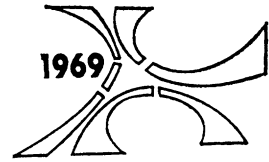


The Importance of the Individual Biological Constitution in the Development of the Various Forms of Cheilo-Gnathopalatoschisis



ALFRED BERNDORFER, M.D.
Budapest, Hungary

The didactic schedule presents several forms of cheilo-gnathopalatoschisis. Clinical practice and observations, however, reveal a considerably larger variety of forms than is commonly described. In the following paper, a thorough investigation of that variety and its embryological and embryopathological interpretation has been attempted.

The fact that one will never find two exactly identical individuals was a basic presumption. Genetic and environmental factors account for the individual differences. The morphological-structural, as well as the biological-biochemical-functional differences have to be investigated in the living subject. The individuality of these differences are commonly pointed out as the "constitution". The constitution is inherent; it develops exactly as the organic structure does. Because of the technical difficulties in human embryology research, the investigation of the biochemical-functional constitution is rather neglected. The problem can be approximated by clinical experience, and by plastic reconstructive surgery.

The body build, as well as physiognomy (if considering only morphology) differs in every single subject. Anthropology records cranial and physiognomic types. If we admit the presence of these anthropological differences in the embryo the fact that these differences evolve during the development has to be acknowledged. They will be present also when some impairment disturbs normal development. The constitutional development proceeds together with the resulting defect. Likewise, the development of the skull, the physiognomy, and the malformation develops, evolves, and takes shape. Accordingly, the malformation should not be considered as an unalterable condition from the moment of its formation. The malformation can undergo changes during the develop-

Dr. Berndorfer is associated with the Paul Heim Pediatric Care Institute, Budapest. This paper was presented at the 1969 International Congress on Cleft Palate, Houston.

ment. Deterioration (even leading to death) may occur; however, repair, regeneration and even complete "restitutio ad integrum" can take place.

According to biological evidence, the earlier the developmental stage (ontogenetic as well as phylogenetic), the higher the restorative ability of the organism. The experiments confirmed that lower organisms will regenerate easily and rapidly after impairment. Given that no such experiments may be carried out in humans, the restorative ability could not be confirmed. Nonetheless, the analysis of single cases and the high number of the varieties recorded are suggestive of the possibility of restoration in the human fetus. The pertinent investigations will be confirmative in case the varieties are set in developmental succession, according to extension and severity of the defect. Should we, for instance, study the cheilo-gnatho-palatoschisis (from the complex and severest defects down to the smallest anomaly) in that developmental succession, we shall find that not only such morphological-mechanical components as severity and size of impairment, the date of occurrence, and its duration, but also the individual disposition, briefly termed as constitution, plays an important part.

Further development is influenced by the malformation; defective organs also affect the development of the other organs. Thus, cheilo-gnatho-palatoschisis should not be considered solely as a local malformation but as a defect of the entire skull including also the face. The variety of the shape of the face and the skull is evident in clinical practice. The degree of the asymmetry of the face and the skull may differ, since this is determined by the formation of the original defect.

Deviation of the nasal septum is present in almost all instances of unilateral clefts, and similarly results in variety. Cheilo-gnatho-palatoschisis should not be considered exclusively as a morphological state. As already mentioned, the living organism consists of an anatomic structure and of a physiological-biochemical function. Structure and function represent an inseparable unit; an impairment to the organism involves simultaneous structural and functional damage. The structure and the function are components of the constitution. Malformations may be considered as a defective constitution.

The tendency to recovery (regeneration) is common in all organisms. The total restoration of the impaired structure and function involves the "restitutio ad integrum"; that is, the subject will be born normal and healthy. Functional defects may, however, remain and appear at some later date of life. In these cases, restoration of the impaired structure or the existence of a morphological defect, not always to be diagnosed in vivo, has to be assessed. Histological examination of the different vital organs is impossible.

There is evidence that the results of reconstructive operations of malformations are not always satisfactory. Sometimes surgery of a large

defect will yield success, whereas unfavorable results are obtained quite often in the operation of minor defects, and the failure cannot always be ascribed to inadequate surgical technique. Here also, constitution may be responsible. If the functional damage is normalized during intrauterine life, the baby will be born and develops, except for a visible defect, as a healthy child with a normal constitution. If, however, the intrauterine regeneration of the defective function was incomplete, the latent defect might become activated by an exogenous effect (such as surgery), and the defective constitution may become manifest. The surgical results will be poor, the children are susceptible to various intercurrent diseases, and they are less responsive to treatment than the normal child.

The malformation should never be considered as a local defect. We do not know the specific cause of a certain malformation. The impairment damages the total organism; it impedes or inhibits development of the organs developing at that time. Different organs develop at the same time. Had the impairment affected all these organs, the child would be born with complex malformations being termed by various syndromes. In the case of a single local defect at birth and no later manifestation of other abnormalities, regeneration of the functional activity of the organism has to be accepted. The conception of an incomplete regeneration is commonly known in experimental embryopathology. The possibility of a partial, incomplete regeneration can be assessed also in the human embryo. The embryologist perceives only a condition but never observes the subsequent development of the defective embryo, if it stays alive. This is one reason why the embryologist, as well as the patho-anatomist, does not see the large varieties that form during the subsequent development.

The clinical studies confirm these theoretical and apparently logical hypotheses of recovery and regeneration process. The large variety of cleft lip, jaw, and palate may be interpreted on the basis of the mentioned biological processes. The diversity may be established by a thorough analysis of the individual cases. Given the deficiency of our present embryological knowledge, the understanding of these varieties requires the simultaneous dynamic study of the structural and functional development.

In numerous cases which have been minutely analyzed, an initial regeneration may be established that points to the tendency of constitutional recovery. In fact, if operations are considered as experiments, we shall find that, in cases we consider to be an incomplete regeneration, the operation will be mostly successful. On the basis of the investigations, it has been concluded that there is an embryological explanation for the variety of malformations and that in addition to the procedure used, success of the operation depends on the biological adjustment of the subject.

Summary

The large number of varieties in cheilo-gnatho-palatoschisis are to be interpreted on the basis of the biological adjustment of the embryo. Each human embryo is an individual organism with specific development. Accordingly, every organism disposes of a regenerative ability to recover from an impairment. This tendency is to be established in numerous forms of malformations, particularly if the different varieties are studied in developmental succession, from the severest defect to the smallest abnormality. In these cases, it is to be seen that not only severity and size of impairment or the date of occurrence and its duration, but also the biological, functional, individual constitution and regeneration ability have to be considered. Since structure and function constitute the living organism, their simultaneous investigation is necessary. The results of operations are dependent also on the individual constitution.

reprints: *Dr. Alfred Berndorfer*
Paul Heim Pediatric Care Institute
Budapest, Hungary

No references presented.