INTRODUCTION

Controversies in the management of single-suture craniosynostosis

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IN clinical practice, the schematic separation of craniosynostoses into simple and complex has constituted the basis for an artificial distinction aimed at facilitating the decision-making process and the management of these malformations. Historically, simple craniosynostoses were grossly identified as the early fusion of one—rarely two—single cranial sutures without associated brain anomalies, while complex craniosynostoses were characterized by the involvement of multiple cranial vault and skull base sutures, associated with underlying alterations of the cerebral structures and CSF spaces and anomalies of the CSF dynamics and cerebrovascular flow. There was a general consensus that the surgical correction of simple craniosynostoses could be limited to reopening the affected suture to allow the growing brain to expand normally, the debate being centered on the age at treatment and choice of the surgical technique. In almost all cases, only one operation was required, which took place during infancy or, less commonly, in childhood. In other words, the management of simple craniosynostoses was rather easy. On the other hand, the corrections of complex craniosynostoses were difficult, requiring a multispecialty team and multiple operations scattered over time due to the progressive nature of the diseases, and involving a multiplicity of morphological and functional anomalies to be cured. Consequently, the action of the surgeon(s) had to be extended during infancy, childhood, adolescence, and even into adulthood, and the corrective surgical procedures were difficult to perform repeatedly.

This rather simplistic schematization is nowadays challenged by the individuation of subtypes of simple craniosynostoses that bear peculiar features and prognoses that require a different cultural and often multispecialty approach, for example, the sagittal synostoses that are part

of specific syndromes (e.g., Sensenbrenner syndrome) or depend on metabolic diseases. Also, the benign outcomes of apparently typical sagittal synostosis are challenged by cases of late recurrent postoperative intracranial hypertension or the development of caudal herniation of the cerebellar tonsils into the upper cervical canal late in life. Trigonocephaly seems to be increasing in incidence in all developed countries, but several cases probably do not need to be operated on as they can normalize during the course of life. Indeed, adults with trigonocephaly are exceptionally or never observed. Consequently, the surgeon should be able to individuate in early life the subjects with premature closure of the metopic suture who should not undergo a surgical correction. The border between simple and complex craniosynostoses may be difficult to individuate in cases of unilateral coronal synostosis, especially type III, that may affect in its progression not only the calvaria but also the skull base and may require the intervention of multiple specialists, namely, ophthalmologists, orthodontists, and craniofacial surgeons. Subtypes of this craniosynostosis have a specific genetic substratum that can interfere with the cognitive development and therefore should be recognized by the pediatric neurosurgeon. Even the impact of the rare lambdoid synostosis is not limited only to the development of the occipital bones but involves also the craniocervical junction and causes various functional impairments and persistent torticollis that respond only partially to the reopening of the affected suture.

In recent years, the introduction of the endoscopic technique has changed the paradigm of our approach to simple craniosynostoses and paved the way to earlier surgical approaches and even interventions limited to the excision of only those segments that appear to be fused in a suture that is not completely closed yet. Controversies have been

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consequently generated on whether these approaches can compete with the traditional open procedures utilized in the neurosurgical practice until now. The papers published in this issue of *Neurosurgical Focus* respond to such a question only in part, but certainly they demonstrate the increased interest as well as the significant impact that the introduction of the newly introduced technical tools has stimulated in the neurosurgical practice.

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