Immediate correction of sagittal synostosis

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John Jane, Milton Edgerton, William Futrell, and Tae Sung Park presented a new technique for the management of sagittal synostosis, described as the $\pi$ procedure, in the Journal of Neurosurgery in 1978. The technique was developed to address the incomplete correction of sagittal synostosis skull deformities seen in patients treated with such techniques as strip and extended strip craniotomy, with or without the supplemental use of a polyethylene film barrier. The strip craniectomy procedure relied on continued normal brain growth to normalize skull shape. It was believed that the incomplete correction of skull deformity was related to a "refusion" of the suture resulting in persistent scaphocephaly. The technique presented by Jane and colleagues differed and represented an important advance because it resulted in immediate correction of skull deformity, rather than relying on brain enlargement as the primary means of remodeling the skull shape. It also was one of the very first to incorporate active reduction of an abnormally long dimension of the skull for the treatment of craniosynostosis. Specifically, in the case of sagittal synostosis, the abnormally long anteroposterior dimension of the skull is shortened by the use of the $\pi$ technique, whereas the abnormally narrowed mediolateral parietal dimension is allowed to expand preferentially by reduced osseous pressure.

The rationale for performing this procedure, as proposed in the original article, when viewed with the advantage of 30 years of additional information-gathering has yielded both support and detraction. Specifically, the authors stated that the likelihood of significant brain maldevelopment as a result of a single-suture craniosynostosis was extremely low; therefore emphasis should be placed on providing appropriate and immediate cosmesis. The psychological benefits of immediate correction of abnormal skull shape to a normal configuration are still very appropriate goals today, even if the results of a number of studies now indicate that a small percentage of patients with sagittal synostosis have increased intracranial pressure and that subtle neural maldevelopment (as evidenced, for example, by learning disabilities) exists with a substantially greater frequency in patients with sagittal synostosis than in children without this disorder. Therefore, the notion of single-suture stenosis as a problem solely of cosmesis is not as strongly supported today as it was in 1978, when the article by Jane et al. was published.

Another issue related to the treatment of craniosynostosis deformity that was addressed in this article was the strip craniectomy extending into the temporal fossa to release the sutural tissue (dura mater) from the skull bone in patent and nonpatent sutures in order to change abnormal dural tensions emanating from the cranial base. This concept does not appear to be central to the thinking regarding craniosynostosis development today. Factors inherent in the dural tissues, such as cytokines, transforming growth factor–$\beta$, and fibroblast growth factor, appear to be playing a more influential role in this issue of resynostosis. The issue of whether there is linkage between dural tension and cytokine expression in craniosynostosis, however, is still being examined.

The $\pi$ procedure did very effectively manage the reshaping of the skull in a single operation, but further refinements in skull shaping followed. These include correction of frontal bossing by remodeling the frontal bone and tilting the remodeled bone posteriorly, expanding narrowed bitemporal diameter, and expanding the diameter of occipital coning.

Finally, the issue of the timing of this operative procedure and its effectiveness at various ages was addressed by the authors. They state that the $\pi$ procedure is most effective in patients younger than 8 months. This recommendation is in keeping with the knowledge that it is easy to reshape immature skull bone, and the potential filling in of bone defects created by surgery at this age is high. In my opinion, however, other techniques are more effective in older children and young adults with mature bone, which is much less malleable and flexible. Techniques such as those described by Marchac and Renier and others involving remodeling of the calvarial vault, appear to be more effective in restoring skull shape to normal at later ages (that is, in patients older than 1 year).

As with any good idea, it is not necessarily the case that the original technique and the rationale for its use are entirely correct when initially described. It is the change in thinking—in this instance, particularly the concept of active remodeling of the skull—that opens doors for further modifications. The $\pi$ procedure has contributed to further investigations into the anatomical pathology of craniosynostosis and theories of skull growth such as the functional matrix theory. This work represents a significant milestone in the thinking regarding the care of patients with sagittal synostosis, which continues to influence thought and the practice of surgery today. (DOI: 10.3171/PED-07/11/426)

References