Organization of Craniofacial Care Dr Linton A. Whitaker Lecture 2023

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Irene M. J. Mathijssen[1]

Dr Linton A. Whitaker is a pioneer of craniofacial surgery. He served as chief of plastic surgery at the Children's Hospital of Philadelphia and University of Pennsylvania. In 2009, he was awarded the Paul Tessier medal by the International Society of Craniofacial Surgery to honor his contributions to the field. Given during the annual meeting of the American Society of Craniofacial Surgeons, the Linton A. Whitaker Lecturer is given by a person whose work or career exemplifies the very best in the art and science of craniofacial surgery.

Since the introduction of craniofacial surgery by Paul Tessier, in collaboration with neurosurgeon Guiot more than 50 years ago, the safety of this type of surgery has improved significantly. This is largely due to better care by the pediatric anesthesiologist, safety of blood transfusions, and to recognition and treatment of associated upper airway anomalies. Imaging of the facial skeleton with CT-scan was of major value. Relevant developments within the surgical field concern the introduction of minimal invasive surgery, distraction osteogenesis and the use of 3D planning.

Providing craniofacial care to children with congenital anomalies of the and skull has significantly improved their quality of life. However, as for many surgical procedures, its effectiveness to improve health of the patient is poorly studied. In the updated version of the guideline on care for craniosynostosis, most included studies were graded with a low level of evidence. This is even the case for basic items of treatment, particularly the indication of surgery in trigonocephaly and for the type and timing of surgery for isolated unisuture, multisuture or syndromic craniosynostosis. Given the big impact that surgery has on the parents and family, we have an obligation to be critical about our performance and we need to scientifically substantiate the proposed procedures.

The majority of publications are descriptive single-center studies that report on retrospectively collected data of a cohort with a low number of patients and a short follow-up time. With the lack of a uniform set of outcome measurements, comparative studies are scarse or they present outcome data that are of less clinical value, for example, surgical time, duration of hospital admittance, costs and blood loss.

The European guideline for craniosynostosis recommends centralization of care for this rare congenital disorder. The minimum volume norm for craniosynostosis per surgeon per year is 20 intracranial procedures. Criteria for the team composition, joint clinics, governance, and available facilities are described. Each center of expertise is obligated to present their activities in a publicly available annual report and be involved in research to further improve care. Also, the recommendation is given to develop standard sets of outcome for each type of craniosynostosis to allow comparative studies between centers for benchmarking and to identify the best treatment protocol.

Within the Netherlands, the process of centralization to 2 centers was undertaken following the first version of the guideline in 2010. At that time, 5 university hospitals provided care for children with craniosynostosis of all types. The total population of the Netherlands is 17.5 million and about 120 newborns with craniosynostosis are born every year. At the end of the process, Erasmus MC in Rotterdam continued the care for all types of craniosynostosis and RadboudUMC in Nijmegen was allocated as second center for isolated single suture craniosynostosis. Both centers are legally bound to deliver care according to the guideline. In accordance with the recommendation in the guideline, the craniofacial center of Erasmus MC presents an annual reports, including all team members and their role within the team, number of surgical procedures and diagnoses, outcomes and complications, and an overview of scientific and educational activities.

Simultaneously, the European Commission developed a strategy to create easy access to specialized care for rare diseases for all European citizens. To achieve this goal, 24 networks were designed, called European Reference Networks or ERNs. One of the 24 ERNs is dedicated to rare craniofacial anomalies and Ear-Nose-Throat disorder, ERN CRANIO. In order to become a member of an ERN, each center had to go through a national procedure to get acknowledgment by its national government as center of expertise for specified conditions. In 2017, all 24 ERNs were established and awarded structural funding by the European Commission. ERN CRANIO is coordinated by me on behalf of Erasmus MC since 2017.

[1]Erasmus Medical Center, Rotterdam, The Netherlands

Corresponding Author:
Irene M. J. Mathijssen, Department of Plastic and Reconstructive Surgery and Hand Surgery, Erasmus Medical Center, Wytemaweg 80, Rotterdam 3015 CN, The Netherlands.
Email: i.mathijssen@erasmusmc.nl

The level of centralization of craniofacial care within Europe differs significantly per member state. About one-third has a clear national policy for centralization with a limited number of centers, one third has no policy at all and one-third has a policy in between. At present, 48 hospitals from 24 member states are member of the ERN CRANIO. The full membership of the craniofacial centers from the United Kingdom changed to supporting partners, given Brexit on January 31st 2020. Patient advocacy groups have prominent role in the network.

The governance within ERN CRANIO is subdivided into 3 workstreams: craniofacial anomalies, cleft lip/palate and orodental disorders, and ear-nose-throat disorders. With the funding supplied by the European Commission, several activities have been deployed: exchange programs between centers for care providers, researchers and patient representatives; development of guidelines (craniosynostosis, craniofacial microsomia, Robin sequence, cleft lip/palate)[1-5] and patient versions of guidelines in all European languages; animations that illustrate surgical procedures for lay person; educational sessions and webinars; the development of a registry to collect outcome data.

Especially the development of a registry is of major importance to improve care for patients. For all included diagnoses with the 24 ERNS, a common dataset is collected, enabling a generalized overview of all rare diseases within Europe. Next to this basic dataset, specific sets of outcomes are defined by the ERN CRANIO working groups for craniosynostosis, craniofacial microsomia, cleft lip/palate and Robin sequence. These data will be collected in the registry and allow multicenter comparative studies, with a much better number of patients to allow research. Given the difference in treatment protocols between centers, these studies will give insight into the impact that various procedures have on outcome.

The tremendous potential of this European network is illustrated with 3 examples.

Example 1. For sagittal suture synostosis, several surgical techniques are available that are performed at various ages. No recommendation could be given within the guideline, based on the available literature on what the best treatment is. For parents, this is one of the most important questions in deciding on treatment for their child. As a first step to answer this question, the working group has developed a photoscore to determine the severity of phenotype through consensus. Testing the created photoscore in a multicenter setting showed a fair to moderate level of agreement between the experts, illustrating the weakness of this tool in general. As a next step, 3D photos are now collected to correlate these data with the 2D photos and refine the photoscore. Also the potential of machine learning will be tested for scoring the 2D photos to improve the reliability. Because not all craniofacial centers have the availability of 3D photogrammetry, analysis of 2D photos will be kept as an outcome measure, to allow all members to contribute to the outcome assessment.

Example 2. Whether or not surgery is indicated for patients with trigonocephaly is under debate. Trigonocephaly is related to a very low risk for raised ICP and a high risk for refraction errors and neurocognitive issues with behavioral problems, despite surgery. The deformity of the forehead also has a natural tendency to improve over time, although it is unknown to what extend this occurs. For trigonocephaly, the working group developed a specific photoscore set to score 2D photos. This set will also be validated with 3D photos because the same fair to moderate level of agreement between experts were detected. Most importantly, a prospective study is undertaken to compare long-term outcome of conservative treatment versus surgery. Initial results of this study indicate that none of the first 100 patients that were treated conservatively developed signs of raised intracranial pressure during the first 4 years of life. Comparing 4-year-old patients that were not operated with patients of similar age that underwent a fronto-orbital advancement (FOA) showed that the first group had slightly more ridging in the midline, but an almost similar shape of the lateral parts of the forehead. The forehead of patients that underwent a FOA was clearly more retruded along its entire width compared to controls of similar age. Comparison at a later age and inclusion of stripcraniectomy plus helmet therapy will follow to establish the added value of surgery.

Example 3. During an annual meeting of the ERN CRANIO, a large variation in postoperative airway management following midface advancement was noticed. This was the start of a multicenter study within our network. Seven centers participated and data of a total of 275 patients with Apert, Crouzon or Pfeiffer syndrome were included. This unique study showed that immediate extubation following midface advancement required no extra respiratory support compared to delayed extubation after a mean of 3 days (range 2-5 days). In addition, immediate extubation was related to a lower number of pneumonia and pressure ulcers compared to delayed extubation. A study with this number of included patients and various treatment protocols is the only way to identify the best protocol to guide future care. Implementation of this recommendation is the next challenging step.

In conclusion, centralization of care for craniosynostosis delivers the required number of procedures to build sufficient expertise and to perform relevant research to further improve care. With the developed outcome sets collected in the European registry, comparative studies can be undertaken to perform benchmarking. It will identify which treatment strategies have the best outcome for our patients and improve their care. The involvement of patients and parents within the network is essential to guide research questions that are important to them and to disseminate knowledge to other patients and parents.

The European model recommends one center of expertise per 10 million inhabitants, a national network in which local hospitals collaborate with the centers of expertise for their mutual patients, and an easily accessible coordinator

within the center of expertise for both patient, parents and local care providers. Care protocols need to be adapted based on solid scientific research and with the input from the patients themselves to bring the level of care to the next level.

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ORCID iD

Irene M. J. Mathijssen https://orcid.org/0000-0002-1675-9922

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