Plastic and Reconstructive Surgery Advance Online Article

DOI: 10.1097/PRS.0000000000003637

Reply: Letter to the Editor: RE: Very low prevalence of intracranial hypertension in trigonocephaly

Martijn J. Cornelissen, MD[1]; Irene M.J. Mathijssen, MD, PhD[1];

1 Department of Plastic and Reconstructive Surgery and Handsurgery, Sophia Children's Hospital, Erasmus University Medical Center.

Corresponding Author

Martijn J Cornelissen

Department of Plastic and Reconstructive Surgery and Handsurgery

Sophia Children's Hospital, Erasmus University Medical Center.

Room Ee15.91, Postbox 2040

3000 CA Rotterdam, the Netherlands

Tel: +31 10 7043292

Fax: +31 10 7044685

Email: m.cornelissen@erasmusmc.nl

Financial disclosure statement

None of the authors has a financial interest in any of the products, devices, or drugs mentioned in this manuscript.

Dear Sir,

We read with interest the reply of Professor Rogers on our paper on the prevalence of intracranial hypertension in trigonocephaly.[1] This paper describes the occurrence of intracranial hypertension, assessed through fundoscopy, and its relation with the occipito-frontal head circumference curve in trigonocephaly patients. This study, in 262 metopic synostosis patients, shows that not only intracranial hypertension (a positive fundoscopy) is rare pre- and post-operatively, but also that it is related to stagnation of the occipito-frontal head circumference curve, as also shown in a previous paper in syndromic craniosynostosis patients.[2]

Professor Rogers raises two concerns with the methodology of the study, on which we are glad to comment. Firstly, the accuracy of fundoscopy in the detection of intracranial hypertension is questioned. As Tuite et al. have shown the sensitivity of fundoscopy in young children may be low.[3] This may have resulted in an underestimation of prevalence of intracranial hypertension in our series, if the patients were assessed by fundoscopy alone. As stated in the discussion section of the paper, we would consider patients with a deflection of the OFC curve, but with a negative fundoscopy, the ones most at risk for a false-negative result of the fundoscopy. These patients underwent repeated fundoscopies and were watched closely for other signs of intracranial hypertension, such as headaches in the morning. We feel that this approach adequately deals with the possible shortcomings of fundoscopy in the screening for intracranial hypertension. Additionally, comparable studies from our center have yielded significantly different results in patients with sagittal and syndromic craniosynostosis of the same age.[2, 4] This, combined with the large number of patients seen in our center, indicates a high level of expertise of our ophthalmologists and a true difference in prevalence of intracranial hypertension in trigonocephaly patients. We feel this cannot be explained by the possible low sensitivity of fundoscopy.

The second issue raised by Prof. Rogers is the diagnostic criteria used to diagnose trigonocephaly, or metopic synostosis. The metopic suture is the first cranial suture to close, mostly within the first year of life. A physiologic closure can indeed occur in the first months of life, without a necessity to operate. True metopic synostosis originates at the 15th week of gestation, resulting in a clear ridge in the midline.[5] In contrast, when the metopic suture has closed at a later stage, some growth has occurred, resulting in a narrow, but not trigonocephalic, forehead with a tipping point bilaterally. This illustrates that the timing of closure is related to the severity of the trigonocephalic shape. We agree with prof. Rogers that a closed metopic suture on itself is not a reason to operate. In our center, the indication for surgery is based on the following assessments: a radiographically confirmed closure of the metopic suture, retrusion of the lateral orbital rim and obvious hypotelorism. The potential bias Prof. Rogers is suggesting is based on two assumptions:

1. Within the patients that were included there is a large group of mild trigonocephaly patients.
2. A mild trigonocephaly results in a lower risk of intracranial hypertension.

In our eyes, these assumptions may not be accurate. Prof. Rogers advocates the use of radiographically-based cranial measurements to standardize the diagnosis and suggests that the use of clinical parameters as mentioned above would lead to wide diagnostic and treatment variability. However, in a paper by Anolik et al., it has been shown that these specific measurements relate closely to the expert decision whether to operate or not.[6] In other words, our clinical judgement would not differ from the computer-based decision whether to operate or not.

Additionally, the prevalence of intracranial hypertension has not been linked to severity of trigonocephaly to date. As previously shown, the intracranial volume of trigonocephaly patients is smaller than controls post-operatively.[7] In contrast, scaphocephaly patients usually

have a normal or larger intracranial volume.[8] Nevertheless, sagittal synostosis patients show a higher prevalence of intracranial hypertension, both pre- and post-operatively.[9] This illustrates that intracranial hypertension is not just a surrogate of cranial shape or 'severity', but is the result of a complex interplay between several parameters, which we may not fully understand to date.

Reference:

1. Cornelissen MJ, Loudon SE, van Doorn FE, Muller RP, van Veelen MC, Mathijssen IM. Very Low Prevalence of Intracranial Hypertension in Trigonocephaly. Plast Reconstr Surg. 2017 Jan;139(1):97e-104e.

2. Spruijt B, Joosten KF, Driessen C, Rizopoulos D, Naus NC, van der Schroeff MP, et al. Algorithm for the Management of Intracranial Hypertension in Children with Syndromic Craniosynostosis. Plast Reconstr Surg. 2015 Aug;136(2):331-40.

3. Tuite GF, Chong WK, Evanson J, Narita A, Taylor D, Harkness WF, et al. The effectiveness of papilledema as an indicator of raised intracranial pressure in children with craniosynostosis. Neurosurgery. 1996 Feb;38(2):272-8.

4. van Veelen ML, Eelkman Rooda OH, de Jong T, Dammers R, van Adrichem LN, Mathijssen IM. Results of early surgery for sagittal suture synostosis: long-term follow-up and the occurrence of raised intracranial pressure. Childs Nerv Syst. 2013 Jun;29(6):997-1005.

5. Mathijssen IM, van Splunder J, Vermeij-Keers C, Pieterman H, de Jong TH, Mooney MP, et al. Tracing craniosynostosis to its developmental stage through bone center displacement. J Craniofac Genet Dev Biol. 1999 Apr-Jun;19(2):57-63.

6. Anolik RA, Allori AC, Pourtaheri N, Rogers GF, Marcus JR. Objective Assessment of the Interfrontal Angle for Severity Grading and Operative Decision-Making in Metopic Synostosis. Plast Reconstr Surg. 2016 May;137(5):1548-55.

7. Maltese G, Tarnow P, Wikberg E, Bernhardt P, Lagerlof JH, Tovetjarn R, et al. Intracranial volume before and after surgical treatment for isolated metopic synostosis. J Craniofac Surg. 2014 Jan;25(1):262-6.

8. Fischer S, Maltese G, Tarnow P, Wikberg E, Bernhardt P, Tovetjarn R, et al. Intracranial volume is normal in infants with sagittal synostosis. J Plast Surg Hand Surg. 2015 Feb;49(1):62-4.

9. Florisson JM, van Veelen ML, Bannink N, van Adrichem LN, van der Meulen JJ, Bartels MC, et al. Papilledema in isolated single-suture craniosynostosis: prevalence and predictive factors. J Craniofac Surg. 2010 Jan;21(1):20-4.