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Original Article

Craniosynostosis incidence with abnormalities of orbital axis on patients under 8 years old

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ABSTRACT

Background: Craniosynostosis may result in malformations of the orbit, which can be observed in clinical presentations. Craniosynostosis impairs the normal growth of the skull, which typically occurs perpendicular to the fused suture. Craniosynostosis is classified into non-syndromic and syndromic, with an incidence of 1: 2000–2500 live births. It is commonly affects the sagittal suture (40–60%), followed by the coronal suture (20–30%), the metopic suture (<10%), and rarely the lambdoid suture. Computed tomography (CT) scan plays a crucial role in identifying the type of cranial abnormality and associated disruptions in the orbital axis (OX).

Methods: The research sample was craniosynostosis patients who were examined at the Radiology Department of Dr. Soetomo General Hospital at Surabaya, Indonesia for the period January 2017–March 2022, male or female aged <8 years and have never had head surgery. Evaluation of the position and axis of the extraocular muscles within the orbits is drawn on the coronal section. In this study, pediatric CT images were acquired at 100 kVp (CTDIvol 2.3 mGy; DLP 84.8 mGy*cm; scan time 6.1 s; helical pitch 0.297). The research was conducted using a case–control method. The case group consisted of patients with craniosynostosis, while the control group included patients without craniosynostosis, encompassing those with conditions such as meningoencephalitis. After the data source is obtained, then the case and control data are matched and then the Chi-square correlation test is carried out through Statistical Package for the Social Sciences.

Results: A significant correlation was found between the incidence of craniosynostosis and abnormalities of OX (*P*-value: 0.000; OR: 22.81; R: 0.635).

Conclusion: There is a significant correlation between the incidence of craniosynostosis that has two or more sutural fusions and abnormalities of OX. Strabismus associated with craniosynostosis is typically detected in patients at an older age. Hopefully, by analyzing the eye angle through CT scans while craniosynostosis is established, abnormalities of the orbital axis can be identified. So the progression of strabismus can be prevented.

Keywords: Computed tomography scan, Craniosynostosis, Cyclorotated, Orbital anomalies, Strabismus

INTRODUCTION

Craniosynostosis is a premature and pathological fusion, either partial or complete, of one or more cranial sutures. This leads to impaired growth of the skull perpendicular to the affected suture. This condition is usually present at birth and can be associated with other anomalies.[1] Primary

craniosynostosis cases occur as an isolated condition in around 85% of cases, while the remaining 15% are associated with multisystem syndrome.[13]

Craniosynostosis is classified into non-syndromic and syndromic with an incidence of 1: 2000–2,500 live births, most commonly in the sagittal suture 40–60%, coronal suture 20–30%, metopic suture <10%, and rarely in the lambdoid suture. The incidence of craniosynostosis syndrome, such as Apert and Crouzon syndrome, is 15.5/million and 16.5/million live births, respectively.[12]

Radiological imaging has an important role in accurate diagnosis, surgical planning, post-treatment evaluation, and identification of coexisting anomalies and complications associated with craniosynostosis.[13] Coexisting anomalies in this case including the orbital aspect, assisting surgical planning, and post-treatment or surgical evaluation. The incidence of craniosynostosis in Indonesia itself has not been well recorded, including there has been no research on the ocular aspects of craniosynostosis, especially in terms of radiological imaging in Indonesia.

Based on this background, researchers were interested in analyzing the correlation of orbital axis (OX) degrees to craniosynostosis, which has been carried out by computed tomography (CT)-Scan Head at Dr. Soetomo General Hospital, Surabaya. Through this research, we hope to detect early changes in OX, allowing for earlier intervention and better management, either surgical or non-surgical.

MATERIALS AND METHODS

The research was conducted after obtaining ethical clearance at Dr. Soetomo General Hospital, Surabaya, Indonesia from January 2017 to December 2022 (ethical number 1281/1184/III/2022). The study was carried out with an observational retrospective approach with a case–control method. The case group consisted of patients with craniosynostosis, while the control group included patients without craniosynostosis, encompassing those with conditions such as meningoencephalitis at the radiology installation of Dr. Soetomo General Hospital for the period January 2017–March 2022. The data sources come from CT-Scan: Toshiba 128 slice type T5× – 101 A, Philips 128 slice type MRC 880, and Siemens 16 slice type M-CT-172 with data in the form of digital raw data. Raw data was measured using software on GE's Picture Archiving and Communication System and Radiant DICOM viewer 2021.2.2. The method used is a manual tracer for each parameter to be measured. The inclusion criteria of this study were male or female craniosynostosis patients aged <8 years and who had never had head surgery. After obtaining the data, the case group and the control group (31:31) were matched, and a Chi-square correlation test was performed using the Statistical Package for the Social Sciences.

It is widely recognized that performing CT scans on children requires a different approach compared to adults. The size of a low-dose CT in children is typically set at around one-half or one-quarter of the dose of a standard head CT scan. The low-dose measurement was conducted according to the study sample, which consisted of patients under the age of 8. Pediatric CT images in this research were acquired at 100 kVp (CTDIvol 2.3 mGy; DLP 84.8 mGy*cm; scan time 6.1 s; helical pitch 0.297). This scanning method aligns with the research conducted by Nagayama *et al*. (2018).[18] This adjustment is made to ensure that the obtained images provide adequate diagnostic information while minimizing radiation exposure and in accordance with as low as reasonably achievable.

Evaluation of the position and axis of the extraocular muscles within the orbit is drawn on the coronal section, with the vertical line intersecting the nasal septum and the horizontal line intersecting the medial and lateral rectus muscles. The angles formed by these axes are then measured in degrees (°), with superior (-) indicating incyclorotation and inferior (+) indicating excyclorotation [Figure 1]. These values are compared to the normal positional range for individuals of the same age.

RESULTS

The data stated that of 31 patients with craniosynostosis, 22 (88.0%) of them experienced an abnormal axis condition. Of the 31 control groups, the majority had a normal OX degree, and only 3 (12.0%) had an abnormal axis condition [Table 1].

The Chi-square test was performed to determine the correlation between the incidence of craniosynostosis and the degree of OX abnormalities. The test results stated that *P* value (Sig 0.000 < 0.005) could mean that there was a significant relationship between the incidence of craniosynostosis and the degree of OX abnormalities [Table 1].

We made several groups based on the degree of ocular tension as follows [Table 2, Figure 1]:

In the case group, if there are at least 2 or more sutures that fused, causing a different degree of OX. For instance, in the normal to mild group, suture closure was present in only one location. In the moderate group, suture closure occurred in two locations. In the severe group, there were more than two complex suture closures [Table 2].

Based on the risk estimate test, the value 22.81 means that the sample with craniosynostosis has a 22.81 times risk of experiencing OX abnormalities compared to control samples without craniosynostosis [Table 3].

The results of the model summary are 0.635 or 63.5%. This value can be stated that the incidence of craniosynostosis affects up to 63.5% of abnormalities in the degree of OX. It can be estimated that out of 100 people with craniosynostosis, 63 of them will experience abnormal OX abnormalities [Table 3].

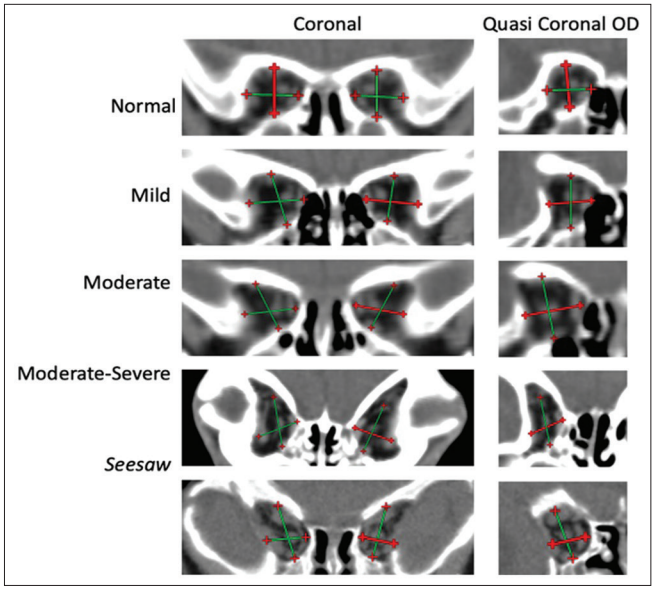

Figure 1: Coronal and quasi coronal computed tomography scan of the posterior orbit of the right occuli. In normal, mild, and moderate criteria, the orbital wall encases the location of m. rectus at its orientation location, in moderate-severe criteria, medial bowing of the greater sphenoid wing (red & green cross arrow) causing m. rectus lateral becomes more inferior, in seesaw (Harlequin deformity), there is a temporal expansion of the orbital roof, indicating a V-pattern, causing lateral displacement of m. rectus superior.[9]

DISCUSSION

Craniosynostosis frequency distribution

Craniosynostosis can lead to papilledema and, ultimately, optic atrophy and vision loss due to elevated intracranial pressure.[7] In non-syndromic craniosynostosis, one study reported papilledema in unicoronal craniosynostosis with prevalence of 3%. In syndromic craniosynostosis, papilledema was most prevalent in Crouzon syndrome 34% [Figure 2] and Apert syndrome 11%.[9,19,20] Thirteen (24%) of 54 patients with various types of craniosynostosis had evidence of optic neuropathy based on fundus examination.[4]

In the functional ophthalmic domain, strabismus was the most prevalent ocular anomaly in both non-syndromic 24% and syndromic craniosynostosis 58%. Strabismus can cause an increased risk of exocyclorotated orbits, which can lead to an incorrect insertion or even malformation of the extra-ocular muscles.[3]

Correlation between craniosynostosis and axis orbita

In measuring ocular torsion, we have to distinguish between subjective torsion and anatomic torsion because these measures can be very different. The one-third disc diameter of the normal torsional range actually subtends about 9° at the fovea. Thus, the torsional positions may differ by 9° from

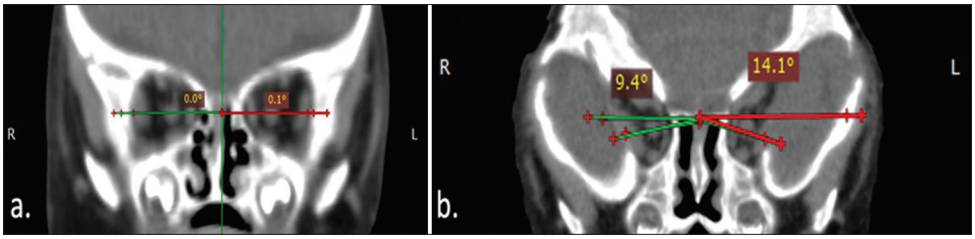

Figure 2: (a) Normal axis in a 7-month-old boy and (b) exyclorotated in a 7-month-old boy with clover leaf skull shape in Crouzon syndrome. The angle formed respresents the intersection of the line between the lateral rectus muscle to the nasal septum and medial rectus muscle to the nasal septum. The green and red lines reveal the position and axis of the extraocular muscle; while the green line represents the right eye and the red line represents the left eye.

Table 1: Craniosynostosis frequency distribution and the correlation between craniosynostosis and axis orbita.

Axis	Cranio		Non-Cranio		Total	
	n	%	*n*	%	*n*	%
Normal	9	24.3	28	75.7	37	100
Abnormal	22	88.0	3	12.0	25	100
Total	31	100	31	100	62	100
	Value	**df**	**Asymp. Sig (2-sided)**	**Exact Sig (2-sided)**		**Exact Sig (1-sided)**
Pearson Chi-square	24.197	1	0.000			
Continuity correction	21.717	1	0.000			
Likelihood ratio	26.550	1	0.000			
Fisher's exact test				0.000		0.000

df: degrees of freedom, Asymp sig: Asymptomatic significance, Exact sig: Exact significance, p value (sig 0.000 < 0.005) stated that there is a strong correlation between the incidence of craniosynostosis and the degree of OX abnormalities

Table 2: Degree of ocular tension and craniosynostosis type.

	Case	Control
Normal (0°–9°)	21	31
Mild (10°–16°)	5	-
Moderate (17°–23°)	4	-
Severe (24°–30°)	1	-
Craniosynostosis Type		
Non syndromic	24	
Syndromic non-specific (facial cleft, achondroplasia, hipotiroid, West syndrome, and Dandy–Walker malformation)	14	
Crouzon syndrome	2	
Apert syndrome	2	
Pfeiffer syndrome	1	

Table 3: Risk estimate and model summary.

			Value	95% Confidence interval	
				Lower	Upper
Odd Ratio for Axis (Normal/Abnormal)			22.815	5.509	94.478
Model	**R**	**R square**	**Adjusted R square**	**Standard error of the estimate**	
1	0.635[a]	0.404	0.384	0.396	

The symbol "a" is the default statistical test result.

those of another and still be labeled within the normal range.[10]

Orbital deformity frequently accompanies the calvarial deformities of craniosynostosis. Elongation and rotation of the vertical axis of the orbital rim are present ipsilateral to coronal synostosis.[10] Compression of the vertical axis with relative widening of the horizontal axis may occur in the orbit contralateral to a unicoronal synostosis (UCS). Widening of the interorbital distance may occur with coronal synostosis, resulting in an ipsilateral increased vertical diameterand tilting of the head as a compensatory mechanism resulting from both the extraocular muscle imbalance.[17] UCS not only affects one coronal suture but also affects orbital skeletal development. There are many disorders, including eyelid anomalies, ptosis and trichiasis, strabismus, proptosis, and refractive error.[8]

The OX is the bisection of the line between medial and lateral orbital walls, while the visual axis is the position of the eye in primary gaze. Both the axis diverges at an angle of 23°. Normally, the equator of the globe is at or slightly anterior to the lateral orbital rim, and the spatial relationship between them is assessed by measuring the distance the globe (top of the cornea) extends over the infraorbital rim, and this distance is generally about 8 mm.[2,11]

Abnormal ocular torsion obviously occurs with palsy or paresis of a cyclovertical muscle. Extorsion occurs with paresis of a superior cyclovertical muscle and intorsion with paresis of an inferior cyclovertical muscle. Because the oblique muscles have greater torsional action than vertical action in the primary position, larger amounts of abnormal torsion are seen with oblique muscle pareses than with rectus muscle pareses. Likewise, oblique muscle surgery, whether a weakening or strengthening procedure, usually has a significant torsional effect.[10]

UCS has effects on ocular motility through the changes in shape and axis of the orbit on the synostotic side. The bony deformation in the frontozygomatic region can result in traction on the ocular globe.[5,6] This direct traction of this region on the lateral check ligament of the lateral rectus muscle causes stretching of the lateral rectus unilaterally in UCS. The stretching results in an increased passive tone of the ocular muscles and an increased extraocular muscle tone from less efficient orbital movements, possibly resulting in strabismus and abnormal extraocular motility. Furthermore, the orbital deformity (Harlequin orbit) results in an abnormal pulley location of the superior oblique, mimicking a weakness of the superior oblique and leading to unopposed action of the inferior oblique muscles, resulting in abnormal extraocular motility and strabismus.[14,15]

In this study, we found some anomaly as brachycephaly, cloverleaf skull, posterior plagiocephaly, scaphocephaly, and other similar cranial deformities. Implementation of the following CT scans in the control group was observed in patients with conditions other than craniosynostosis. These conditions included meningoencephalitis. Using some control group of patients, the patients are inevitably young, and therefore, it is difficult to diagnose and objectify ophthalmic sequelae, possibly leading to an underestimation of the prevalence of the described complications preoperatively.

Therefore, it remains important to keep young patients with UCS under precise orthoptic and ophthalmologic examinations, independent of the severity of UCS. Small manifest squints have the same inherent effect of visual loss through amblyopia as larger squints. Clinicians need to be aware that this can occur on the non-synostotic side as well as the synostotic side. The patient should be kept under close regular monitoring both pre-and postoperatively.[18]

Axial CT scanning augmented with coronal and longitudinal orbital reformatted images has been useful for quantitative spatial localization of the globes. Nonetheless, the slice format of these images requires imagination for surgical planning.[16]

CONCLUSION

The diagnosis of craniosynostosis is based on clinical features and investigations. Radiological examination is important for making an accurate diagnosis, surgery planning, evaluating therapy, and identifying accompanying anomalies and complications related to craniosynostosis surgery. This study found that there is a significant correlation between

craniosynostosis and abnormalities of OX. Strabismus associated with craniosynostosis is typically detected in patients at an older age. By analyzing the OX through CT scans during the diagnosis of craniosynostosis, earlier diagnosis and treatment can be achieved, potentially preventing the progression of strabismus.

Ethical approval

The research/study approved by the Institutional Review Board at Dr. Soetomo General and Academic Hospital, number 1281/1184/III/2022, dated 2022.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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