CT-SCAN IMAGES OF THE POSTERIOR ORBITAL, ORBITAL APEX, EXTRAOCULAR MUSCLES POSITION AND AXIS IN CRANIOSYNOSTOSIS

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ABSTRACT

Introduction: Craniosynostosis is the premature and pathological fusion, either partial or complete, of one or more cranial sutures. One of the ophthalmological complications is strabismus (in 39–76% of cases), with more severe manifestations in syndromic craniosynostosis. Head-CT with 3D-reconstruction can be used to evaluate the anatomical abnormalities of the orbital aspects to assist clinicians in planning surgery.

Materials and Methods: A retrospective descriptive–observational study was conducted for 43 patients in the period from January 2017 to March 2022. The patients had syndromic or non-syndromic craniosynostosis and underwent head CT-scans at Dr. Soetomo General Hospital, Surabaya. The posterior orbital bones, orbital apexes, positions and axes of the extraocular muscles were evaluated by an neuroimaging radiology consultant.

Results: 26 male subjects (60.5%) and 17 female subjects (39.5%) were evaluated. 18 patients (41.9%) had normal posterior orbital configurations and extraocular muscles, 4 (9.3%) showed mild abnormalities, 13 (30.2%) had moderate abnormalities, 2 (4.7%) showed moderate–severe abnormalities, and 6 (11.6%) had seesaw pattern abnormalities. 33 patients (76.7%) had normal orbital apex configurations, 3 (7%) showed ballooning ethmoidal sinuses, 2 (4.7%) had protrusions of the lateral orbital wall (greater sphenoid wing), and 5 (11.6%) showed combined ethmoidal sinus ballooning and protrusion of the lateral orbital wall.

Conclusion: The smallest excyclorotation was observed for the mild abnormalities of the posterior orbital configuration and extraocular muscles in the right eye (4.5–11.3°) and left eye (3.6–7°). The greatest was noted for the seesaw pattern in the right eye (7.6–21.4°) and left eye (7.4–30°). 3 patients (7%) had an incyclorotated axis and 26 (60.5%) had an excyclorotated axis, with a greater degree of excyclorotation in the seesaw pattern.

Keywords: craniosynostosis, orbital, excyclorotated, strabismus, ct-scan

INTRODUCTION

Craniosynostosis is the premature and pathological fusion, either partial or complete, of one or more cranial sutures. It is defined as single if it involves only one suture and is said to be complex if it involves multiple sutures.[1] Craniosynostosis is classified as either syndromic or non-syndromic, with an incidence of 1:2000–2500 live births. It occurs most commonly in the sagittal suture (40–60%) or coronal suture (20–30%), occasionally in the metopic suture (less than 10% of cases), and rarely in the lambdoid suture.[2] The incidence of syndromic craniosynostosis, most commonly Apert and Crouzon syndromes, is 15.5:1,000,000 and 16.5:1,000,000 births, respectively.[3,4]

The most common ophthalmological complication of craniosynostosis is optic neuropathy, which results from increased intracranial pressure in 30–40% of syndromic and in 15–20% of non-syndromic cases.[5] Strabismus occurs in 39–76% of cases with pathogenesis related to the abnormal orbital shape and impaired function of a number of extraocular muscles. Astigmatism and anisometropia occur in 40% and 16% of these pathogenesis cases, respectively, and exposure keratopathy may occur due to exorbitism. Amblyopia is caused by one or more of the above conditions.[6,7] Ophthalmologic problems are more frequent and more severe in syndromic craniosynostosis.[8] When the underlying cause of amblyopia is not detected and treated early, the cortical development window may close and visual disturbances may become permanent.[7]

Head CT scans and 3D reconstruction are the primary imaging modalities for evaluating the anatomical aspects of the orbit in craniosynostosis.[9] The incidence of craniosynostosis, including complications often associated with this disorder, is not well recorded in Indonesia.[10] Until now, no research has been conducted in Indonesia regarding the ocular aspects of craniosynostosis, especially using imaging.

Based on this background, researchers are interested in studying CT-scan images of the posterior orbital bone, orbital apex, position and extraocular muscle axis in patients with craniosynostosis. It is hoped that radiology could improve accuracy in the diagnosis of craniosynostosis and its manifestations and complications in the orbit to assist clinicians in determining treatment and assessing the prognosis, in this case relating to the visual function of craniosynostosis.

RESEARCH METHODS

This research was conducted through an observational, descriptive, retrospective approach from January 2017 to March 2022 in syndromic and non-syndromic craniosynostosis patients who had undergone head CT scans at Dr. Soetomo General Hospital in Surabaya. Inclusion criteria: male or female patients aged less than 8 years. Exclusion criteria: history of trauma and surgery in the head area. Head CT scans were performed using a Toshiba 128-slice type T5x – 101 A, Philips 128-slice type MRC 880 and Siemens 16-slice type M-CT-172. Data were obtained in the form of digital raw data and were evaluated using GE's Picture Archiving and Communication System (PACS) and Radiant Dicom viewer 2021 (version 2.2) software.

Evaluation of the configuration of the posterior orbital bone, orbital apex, and position and axis of the extraocular muscles was conducted by a neutral observer (neuro-radiologist). Classification of the degrees of severity of the posterior orbital bone configuration, orbital apex and position of the extraocular muscles was conducted according to the criteria set out by Dagi et al., 2017. The axes of the horizontal (medial and lateral) rectus muscles were measured using a vertical line on the nasal septum and a perpendicular horizontal line. The axes were then determined to be excyclorotated or incyclorotated through data analysis with MS Excel and SPSS 25.0 software.

RESULTS

The research sample of 43 patients was taken from the total eligible population (consecutive sampling).

Table 1. Subject characteristics

Variable	Value [n(%)]
Number of Subjects	43
Age (in months)	
Average ± Standard Deviation	9.12 ± 11.85
Median	5
Range	1–63
Gender	
Male	26 (60.5%)
Female	17 (39.5%)
Craniosynostosis	
Non-Syndromic	
- Multiple	22 (48.8%)
- Single (metopic, sagittal, squamosal)	3 (7%)
Syndrome	19 (44.2%)
- Apert	2 (4.6%)
- Crouzon	3 (6.9%)
- Pfeiffer	1 (2.32%)
- Facial cleft (non-specific)	5 (11.6%)
- Dandy–Walker malformation (non-specific)	2 (4.64%)
- Achondroplasia (non-specific)	1 (2.32%)
- West syndrome (non-specific)	1 (2.32%)
- Non-specific (unknown)	4 (9.28%)
Closing Sutures	
Bicoronal	6 (14.3%)
Bicoronal, sagittal	1 (2.3%)

Bicoronal, bisquamosal	10 (23.3%)
Bicoronal, bilambdoidea	1 (2.3%)
Bicoronal, bilambdoidea, sagittal	2 (4.7%)
Bicoronal, left squamosal, left lambdoidea	1 (2.3%)
Bicoronal, bisquamosal, right lambdoidea	3 (7%)
Bicoronal, bisquamosal, left lambdoidea	1 (2.3%)
Bicoronal, bisquamosal, bilambdoidea	6 (14%)
Bicoronal, bisquamosal, bilambdoidea, sagittal	7 (16.3)
Bicoronal, bisquamosal, sagittal, right lambdoidea	1 (2.3%)
Metopic, bicoronal, right lambdoid	1 (2.3%)
Sagittal	1 (2.3%)
Metopic	1 (2.3%)
Left Squamosal	1 (2.3%)
Configuration of the Posterior and Extraocular Muscle Bones	
Normal	18 (41.9%)
Mild	4 (9.3%)
Moderate	13 (30.2%)
Moderate–Severe	2 (4.7%)
Seesaw	6 (11.6%)
Apex Orbital Configuration	
Normal	33 (76.7%)
Ballooning ethmoidal sinus	3 (7%)
Protrusion of the lateral orbital wall (greater sphenoid wing)	2 (4.7%)
Ballooning ethmoidal sinus and protrusion of the lateral orbital wall (greater sphenoid wing)	5 (11.6%)
Right and Left Eye Axis	
Normal	14 (32.6%)
Incyclorotated	3 (7%)
Excyclorotated	26 (60.5%)

26 of the patients in this study (60.5%) were male. The mean age in this study was 9.12 ± 11.85 months.

24 patients (55.8%) had non-syndromic craniosynostosis. Of these, 22 (51.2%) had multiple synostosis and 2 (4.7%) had single synostosis. 19 patients had syndromic craniosynostosis (44.2%). Most suture closures (in 10 patients; 23.3%) were bicoronal bisquamosal. 18 patients (41.9%) had normal posterior bone and extraocular muscle configurations, 4 (9.3%) showed mild abnormalities, 13 (30.2%) had moderate abnormalities, 2 (4.7%) showed moderate–severe abnormalities, and 6 (11.6%) had a seesaw configuration. 33 patients (76.7%) had a normal orbital apex configuration, 3 (7%) showed

ballooning ethmoidal sinus, 2 (4.7%) had protrusion of the lateral orbital wall (greater sphenoid wing), and ballooning ethmoidal sinus lateral and orbital wall protrusion (greater sphenoid wing) was observed in 5 patients (11.6%). The right and left eye axis was normal in 14 patients (32.6%), incyclorotated in 3 (7%), and excyclorotated in 26 (60.5%).

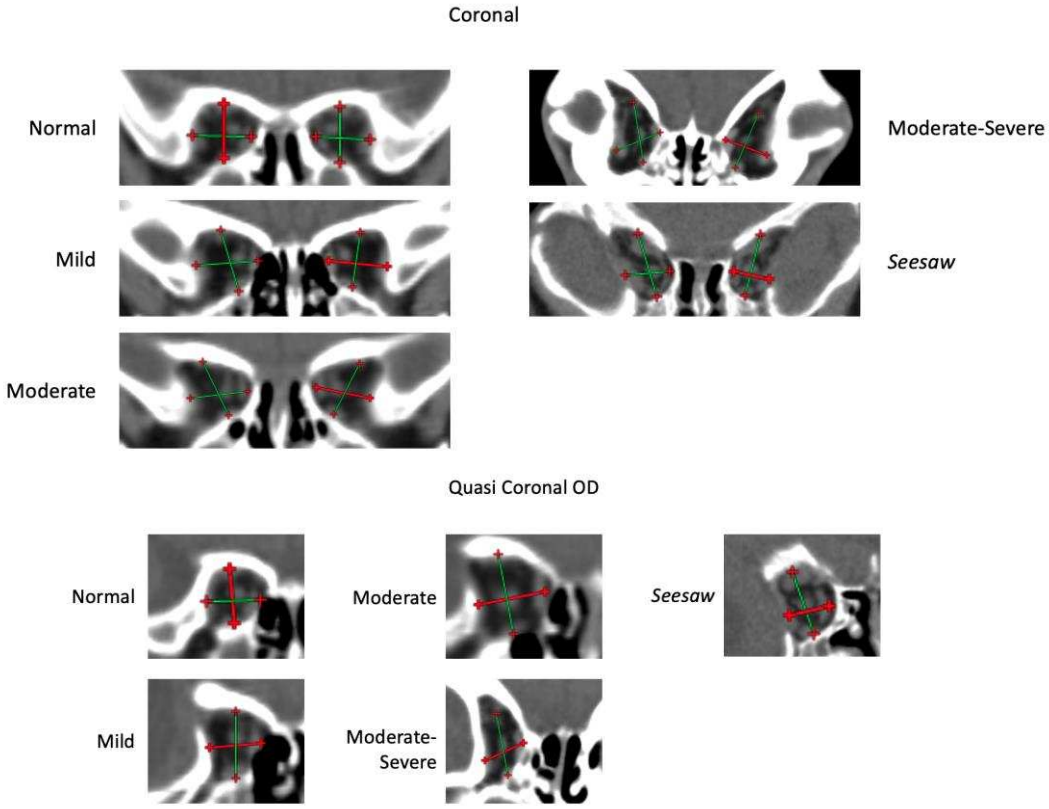

Figure 1. Coronal and quasi-coronal oculi dextra posterior orbit CT-scans. The criterion for mild and moderate abnormalities is that the orbital wall encases the rectus muscle in its local orientation. In moderate–severe cases, medial bowing of the greater sphenoid wing (yellow arrow), causes the lateral rectus muscle to become inferior. The seesaw conformation (harlequin deformity) involves temporal expansion of the orbital roof, indicating a V-pattern, causing lateral displacement of the superior rectus muscle.

100% of patients with syndromic craniosynostosis achondroplasia, Dandy–Walker malformation, facial cleft and Pfeiffer syndrome had normal orbital apex configurations. 100% of patients with Apert syndrome had ballooning ethmoidal sinuses and protrusions of the lateral orbital wall.

33.3% of patients with Crouzon syndrome had ethmoidal sinus ballooning, and 66.7% had a combination of ballooning ethmoidal sinus and protrusion of the lateral orbital wall. 75% of patients with non-specific syndromic craniosynostosis had a normal orbital apex configuration, and 25% had a combination of ballooning ethmoidal sinus and protrusion of the lateral orbital wall.

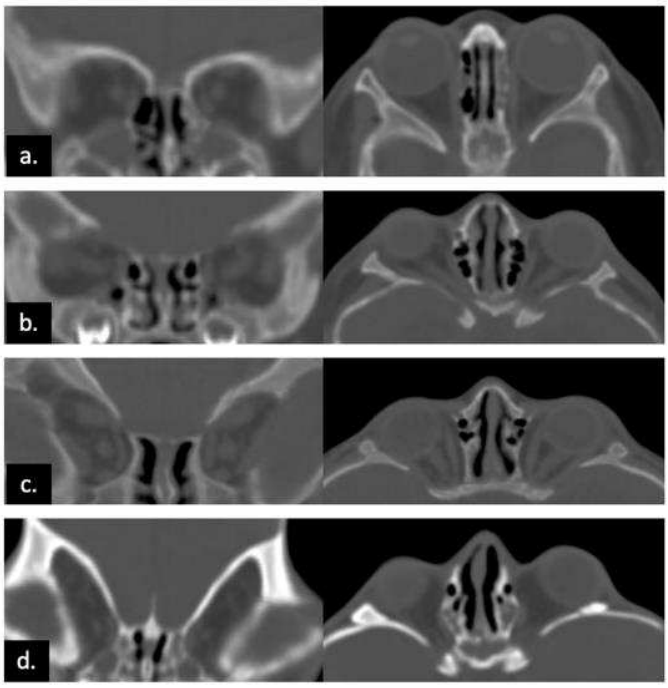

Figure 2. Coronal and axial CT scans of the orbital apex: (a) normal, (b) ethmoidal sinus ballooning, (c) orbital lateral wall protrusion, (d) combined ethmoidal sinus ballooning and orbital lateral wall protrusion. (b) and (d) occur in Apert and Crouzon syndromic craniosynostosis.

100% of patients with achondroplasia or West syndromic craniosynostosis had a normal posterior bone configuration and extraocular muscles. As many as 100% of patients with Apert or Pfeiffer syndrome had a seesaw pattern posterior bone configuration and extraocular muscles. 33% of Crouzon syndrome patients has a moderate configuration of posterior bone and extraocular muscles, and 66.7% showed a seesaw pattern. For patients with syndromic craniosynostosis with Dandy–Walker malformation, 50%

had a normal posterior bone configuration and 50% had moderate extraocular muscles. As many as 60% of patients with non-specific (unknown) syndromic craniosynostosis and 40% of patients with facial cleft had a normal posterior bone configuration and moderate extraocular muscles. The explanation of this percentage is summarised in Table 2.

Table 2. Percentage of syndromic craniosynostosis patients (each type) with different classifications of posterior bone configuration and extraocular muscles

Syndromic	Posterior bone configuration and extraocular muscles				
	Normal	Light	Current	Medium–Heavy	Seesaw
Apert	0.0%	0.0%	0.0%	0.0%	100.0%
Crouzon	0.0%	0.0%	33.3%	0.0%	66.7%
Pfeiffer	0.0%	0.0%	0.0%	0.0%	100.0%
Dandy–Walker malformation	50.0%	0.0%	50.0%	0.0%	0.0%
Facial cleft	60.0%	0.0%	40.0%	0.0%	0.0%
Achondroplasia	100.0%	0.0%	0.0%	0.0%	0.0%
West syndrome	100.0%	0.0%	0.0%	0.0%	0.0%
Non-specific (unknown)	60.0%	0.0%	40.0%	0.0%	0.0%

Table 3. Percentage of posterior orbital bone configuration and extraocular muscles, with extraocular muscle axes (right and left)

Posterior bone configuration and extraocular muscles	Incyclorotated Extraocular Muscle Axis		Excyclorotated Extraocular Muscle Axis	
	Right	Left	Right	Left
Normal	$2.7–10.6^0$	$9.7–10^0$	$0–1.4^0$	$0–5.3^0$
Mild	-	-	$4.5–11.3^0$	$3.6–7^0$
Moderate	-	-	$3.3–14.7^0$	$4.2–18.9^0$
Moderate-Severe	-	-	$6.1–23.5^0$	$6.9–20.3^0$
Seesaw	7^0	18^0	$7.6–21.4^0$	$7.4–30^0$

Table 4. Configuration of the posterior orbital bones and extraocular muscles with axes of extraocular muscles (right and left)

Posterior bone configuration and extraocular muscles	Extraocular Muscle Axis (Right & Left)		
	Normal	Incyclorotated	Excyclorotated
Normal	77.8%	11.1%	11.1%
Mild	0.0%	0.0%	100.0%
Moderate	0.0%	0.0%	100.0%
Moderate–Severe	0.0%	0.0%	100.0%
Seesaw	0.0%	16.7%	83.3%

100% of patients with a posterior orbital bone configuration and normal and mild abnormalities in extraocular muscles had a normal orbital apex configuration. As many as 84.6% of patients with posterior orbital bone configuration and extraocular muscles with moderate abnormalities had a normal orbital apex configuration. 50% of patients with posterior orbital bone configuration and extraocular muscles with moderate to severe abnormalities had an orbital apex configuration with ballooning ethmoidal sinus and lateral orbital wall protrusion (greater sphenoid wing). Of the patients with posterior orbital bone configuration and seesaw patterned extraocular muscles, 66.7% had orbital apex configuration with ballooning ethmoidal sinus and lateral orbital wall protrusion (greater sphenoid wing).

DISCUSSION

Most of the 43 patients were male (26; 60.5%) and 17 (39.5%) were female. A previous study in Asia by Byun et al. had a gender ratio of 59% male and 40.9% female.[11] Research in America by Gonzales et al. included 64.5% male and 34.5% female patients.[12] Until now, no research has explained the reasons for male predominance in the patient samples.

There were 24 patients with non-syndromic craniosynostosis (55.8%), 22 (51.2%) had multiple disorders, and 2 (4.7%) had single synostosis. This is in contrast to the study by Byun et al., in which 17.2% of patients had multiple types of non-syndromic craniosynostosis and 72.59% had single synostosis.[11] It also differs from the study by K. Senarath-Yapa et al., in which the proportions of patients with multiple non-syndromic craniosynostosis and single synostosis were 5–15% and 85–95%,

respectively.[13] While 19 patients (44.2%) had syndromic craniosynostosis, this was also different from other studies, for instance research by Tonne et al. in which 27% of patients had syndromic craniosynostosis.[14]

Considering the abnormalities of multiple sutures, as many as 10 (23.3%) were due to the closure of bicoronal sutures. This is consistent with previous studies in which the bicoronal suture was the most common type associated with multiple craniosynostosis, both syndromic and non-syndromic.[11,14]

The grouping of the orbital aspects in this study follows previous research conducted by Dagi et al., who observed the influence of the shape of the posterior orbital aspect on the position and the extraocular muscle axis in craniosynostosis with excyclotorsion strabismus. Classic strabismus craniosynostosis has a characteristic “excyclotorsional syndrome”.[6,15,7] In coronal craniosynostosis there is flattening of the fronto-parietal region, which causes the antero-posterior diameter of the orbital cavity to shorten, widening the palpebral fissure on the affected side. If severe enough, a harlequin sign will appear. In previous studies by Dagi et al., the most severe form is the seesaw pattern.[15,16] In syndromic craniosynostosis there will also be anterior protrusion of the lateral orbital wall, sphenoid greater wing and lateral ballooning of the ethmoidal sinus.[15] The more severe the category of posterior bone configuration abnormalities and position of the extraocular muscles, the greater the degree of the excyclorotated axis.[15]

In this study, 100% of patients with Apert or Pfeiffer syndromes had posterior bone configuration and an extraocular muscle seesaw pattern.

33.3% of patients with Crouzon syndrome had moderate posterior bone configuration, and 66.7% had seesaw extraocular muscle abnormalities. A study by Dagi et al. identified a significant relationship between Apert syndrome and moderate–severe or seesaw V-Pattern strabismus, and between Crouzon or Pfeiffer syndrome and mild to moderate V patterns.[15] The genes that are often mutated in craniosynostosis are in the FGFR family because of their high expression at the sutures.[17] FGFR2 is the most widely distributed throughout the sutures, mutating most frequently and affecting the coronal, metopic, sagittal and lambdoid sutures. The FGFR3 mutation only affects the coronal and metopic sutures, whereas FGFR1 (as well as Twist1 and EFNB1) usually affects only the coronal suture.[18] Apert syndrome is characterised by a dominant FGFR2 mutation; the mutation is in FGFR2 and FGFR3 for Crouzon syndrome, and FGFR1 and FGFR2 for Pfeiffer syndrome. Achondroplasia is characterised by an FGFR 3 mutation, while there are currently no studies on craniosynostosis in facial cleft and West syndrome.

100% of patients with Apert syndrome had an orbital apex configuration with ballooning ethmoidal sinuses and protrusion of the lateral orbital wall. The high incidence of seesaw patterns and morphological abnormalities of the orbital apex (ballooning ethmoidal sinus and protrusion of the lateral orbital wall) for Apert syndrome may be related to earlier occurrences of craniosynostosis, such as the closing of a coronal suture at birth with sagittal and metopic sutures.[19] Patients with Crouzon syndrome may present as normal at birth, but craniosynostosis will occur during the first two years.[19]

CONCLUSIONS AND RECOMMENDATIONS

18 patients (41.9%) had craniosynostosis with a normal configuration of the posterior bone and extraocular muscles. 4 (9.3%) had mild abnormalities, 13 (30.2%) had moderate abnormalities, 2 (4.7%) showed moderate–severe abnormalities, and 6 (11.6%) had a seesaw configuration. 33 patients (76.7%) had craniosynostosis with normal orbital apex configuration, 3 (7%) showed ballooning ethmoidal sinus, 2 (4.7%) showed protrusion of the lateral orbital wall (greater sphenoid wing), and 5 (11.6%) had a combination of ballooning ethmoidal sinus and lateral wall protrusion orbit.

The smallest excyclorotation occurred for mild abnormalities in the posterior orbital and extraocular muscle configurations in the right eye (4.5–11.3°) and left eye (3.6–7°). The largest was observed for the seesaw pattern in the right eye (7.6–21.4°) and the left eye (7.4–30°). 3 patients (7%) had incyclorotated right and left eye axes were. These were excyclorotated in 26 patients (60.5%), with a greater degree of excyclorotation in the posterior bone configuration and more severe muscle abnormalities.

Further research is needed to determine the descriptions and relationships of the posterior orbital bone configuration, orbital apex, and muscle position and axis. We suggest studying extraocular patients without craniosynostosis abnormalities to generalise the results and understand the differences for craniosynostosis patients.

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