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Craniofacial Deformities: Original Research

Three-Dimensional Computed Tomography Imaging in the Evaluation of the Child With Craniosynostosis: Study of 60 Patients*

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ABOUT ARTICLE

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ABSTRACT

Purpose. The aim of this article is to describe and illustrate the appearance of craniosynostosis on CT scan images.
Methods. A total of 60 children during the years 2014-2018 were included. The medical and radiographic records of patients less than 3 years of age, clinically diagnosed with nonsyndromic craniosynostosis were studied. They underwent CT imaging examination in order to confirm the diagnosis and to accurately establish the surgical treatment plan.
Results. The high diagnostic performance of CT with 3D surface-rendered reconstructions was pointed out together with the most common findings.
Conclusion. 3D CT appears as the imaging modality with the best diagnostic performance in children with craniosynostosis. However, large prospective studies and further research are needed, in order to clearly define the role of 3D CT and minimize the unnecessary exposure of infants to radiation.

"CT imaging is currently considered the criterion standard for diagnosing craniosynostosis."
—Jeffrey A. Fearon, 2012[1]
USA

Craniosynostosis is an important clinical entity in the pediatric population resulting in significant cosmetic and functional effects. This pathological condition of infancy, characterized by partial or complete premature fusion of one or multiple cranial sutures, leads to characteristic skull shape deformities and facial asymmetry.[1] It occurs in approximately 3.5–4.5 out of 10,000 live births worldwide. It can affect one or multiple sutures, occur as an isolated defect or be associated with a craniofacial syndrome. The incidence of non syndromic cases is higher compared to syndromic.[2]

Premature fusion of a suture leads to growth restriction of the skull perpendicular to the suture`s axis. This results in characteristic anomalies of skull shape, which can be readily diagnosed clinically and with imaging. The main causes of morbidity in craniosynostosis are increased intracranial pressure, headaches, neurodevelopmental delay, visual defects and cosmetic deformities.[3]

Although the diagnosis of craniosynostosis can be clinical, all imaging techniques contribute to the accurate diagnosis of the entity. The overall goal of neuroimaging for infants with craniosynostosis is the early detection and description of this entity to enable appropriate treatment.[4] Delayed diagnosis and treatment may lead to cosmetic calvarial deformity which may be difficult to correct, or may require more extensive cranial reconstruction. In addition, may cause a potentially irreversible neurological impairment. Specific imaging aims include detailed characterization of the number of sutures, exact extent of the suture fusion, demonstration of the resulting

* This manuscript has not been presented

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craniofacial deformities, as well as the underlying structural brain changes and the coexisting anomalies or complications.[5]

Classically, plain skull radiography is the initial technique used in the evaluation of a child with an abnormal head shape due to its greater availability, low cost and reduced risk.[6] Three-dimensional computed tomography (3D CT) is considered the gold standard for the diagnosis of craniosynostosis in the assessment of infants with abnormal skull shape, as it can provide highly detailed 3D images of the skull, along with information about the possible coexisting anomalies of the brain.[7]

As a rule, 3D CT is the imaging modality of choice for syndromic cases, provided that the number and extent of suture fusion are described in detail.[8] On the other hand, for nonsyndromic cases, the suggested diagnostic algorithm includes plain radiographs for the initial assessment, followed by 3D CT only to establish the complex surgical treatment plan. With all this theoretical background, the objective of the current article is to present the characteristic radiologic features of craniosynostosis on 3D CT imaging.

Methods

At the Department of Pediatric Surgery, Division of Pediatric Craniofacial Surgery at the National Institute of Children Diseases, a total of 60 children (36 males and 24 females) with nonsyndromic craniosynostosis have been monitored by 3D CT while creating the treatment plan according to the guidelines of the World Medical Association (Declaration of Helsinki). In addition, a standard measurement protocol, examined and approved by the Institutional Ethical Committee was used.

The average age at the time of first presentation was 5.2 months, with a range between 3.2 and 9.4 months (Chart 1). All patients underwent primary surgery with open cranial vault reconstruction, with the minimum requirement for 3D CT examination preoperatively to confirm the diagnosis and to create the ideal surgical protocol. Evaluation of each case was performed clinically and radiographically by the author in consultation with a pediatric neuroradiologist. Every CT scan was examined carefully and all characteristic radiographic features were pointed out in addition to the clinical picture of each child.

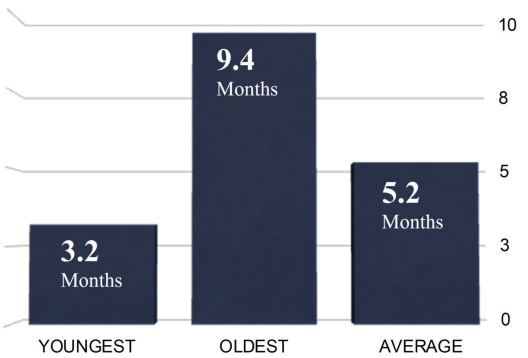

CHART 1. Distribution of age (months) at the time of diagnosis.

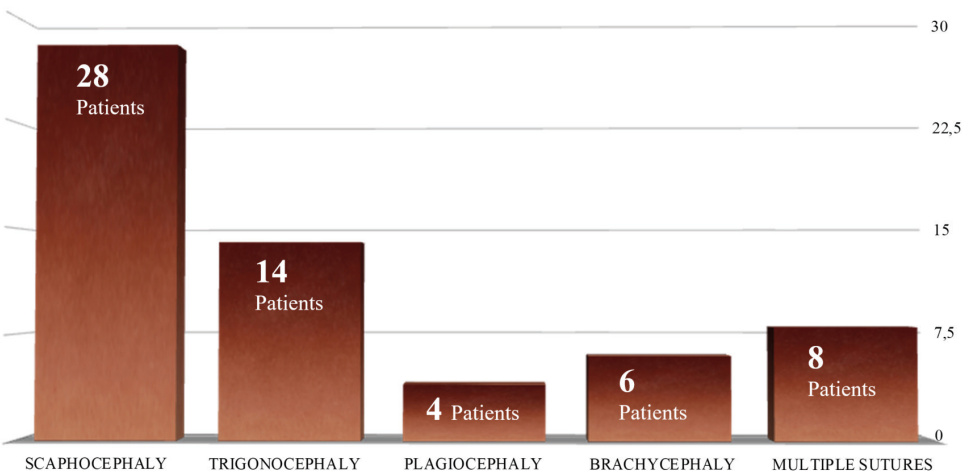

CHART 2. Distribution of patients with head shape deformity as a result of craniosynostosis.

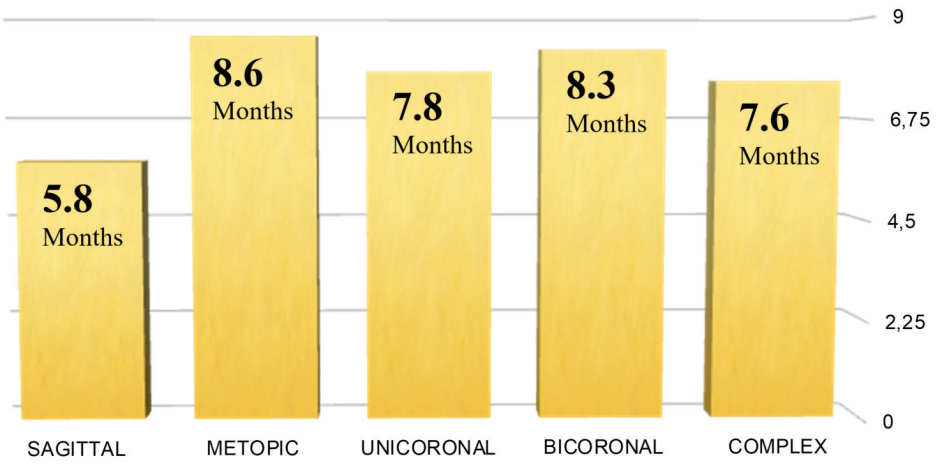

CHART 3. Average age (months) at the time of diagnosis in relation to the synostotic suture.

Results

Sagittal synostosis was the most common synostosis and included approximately 50 percent of our cases (Chart 2). The mean age at the time of first presentation was 5.8 months (Chart 3). Fusion of the sagittal suture restricts growth in the transverse dimension (perpendicular to the suture axis), with compensatory overgrowth in the anteroposterior dimension (along the suture axis). This resulted in a narrow elongated skull with increased AP dimension and decreased transverse dimension (Fig 1A-D). Prominent frontal and occipital protuberances as well as ridging at the suture were seen. The term dolichocephaly describes the skull's shape after the early synostosis of the sagittal suture. Scaphocephaly is a subgroup of dolichocephaly, including evident ridging of the sagittal suture, alike to the boat's keel.

Fourteen children demonstrated a premature closure of the metopic suture at a mean age of 8.6 months at the time of first consultation. Premature fusion of the metopic suture results in symmetrical bone growth at the sagittal suture and asymmetric bone growth at the coronal sutures. This produced a characteristic triangular or pear-shaped frontal deformity called trigonocephaly (Fig 2A-D). Trigonocephaly corresponds to the resulting deformity from synostosis of either the metopic suture (anterior trigonocephaly), or the posterior third of the sagittal suture and both the lambdoid sutures (posterior trigonocephaly). Associated findings included hypotelorism, parietooccipital bossing, narrow anterior cranial fossa, lateral orbital hypoplasia, characteristic metopic notch noted on the inner side along the suture line, metopic ridge on the outer side of the suture, hypoplastic ethmoid sinuses, deficient supraorbital ridges, and a medial upward slanting of the orbital roofs.

The 4 children enrolled in the study with premature unilateral coronal synostosis had a median age of 7.8 months at the time of first meeting. Unilateral coronal synostosis caused restricted growth in the AP dimension and compensatory overgrowth in the transverse dimension, which resulted in anterior plagiocephally over the fused suture, with ipsilateral temporal expansion and contralateral frontal and parietal expansion (Fig 3A-D). Associated findings included shallow anterior cranial fossa, shallow orbits, depression of the petrous bone, and the "harlequin eye deformity" (upward slanting of the orbital roof laterally).

Six children with premature bilateral coronal suture synostosis were included in our series with a mean age of 8.3 months at the time of first evaluation. Bilateral coronal synostosis resulted in decreased AP dimension of the calvaria, which is called brachycephaly (Fig 4A-D). An abnormally tall head (turricephaly) results from symmetrical bone growth along the sagittal and metopic sutures. Bicoronal synostosis is often associated with other synostoses and malformation syndromes.

Eight children with multiple sutures` synostosis presented to our department. The average age during first examination was 7.6 months. Four patients demonstrated a fusion of the unilateral coronal suture in association with the sagittal suture (Fig 5A-D). Three patients presented with fusion of the metopic and sagittal sutures and one patient encountered fusion of the metopic suture together with the coronal sutures bilaterally. Complex craniosynostosis usually occurs in syndromic cases. Deformities depend on varying combinations of sutures involved. Nevertheless, in cases of combined synostosis of all major sutures, the skull has a cloverleaf appearance, which is the most impressive craniofacial deformity.

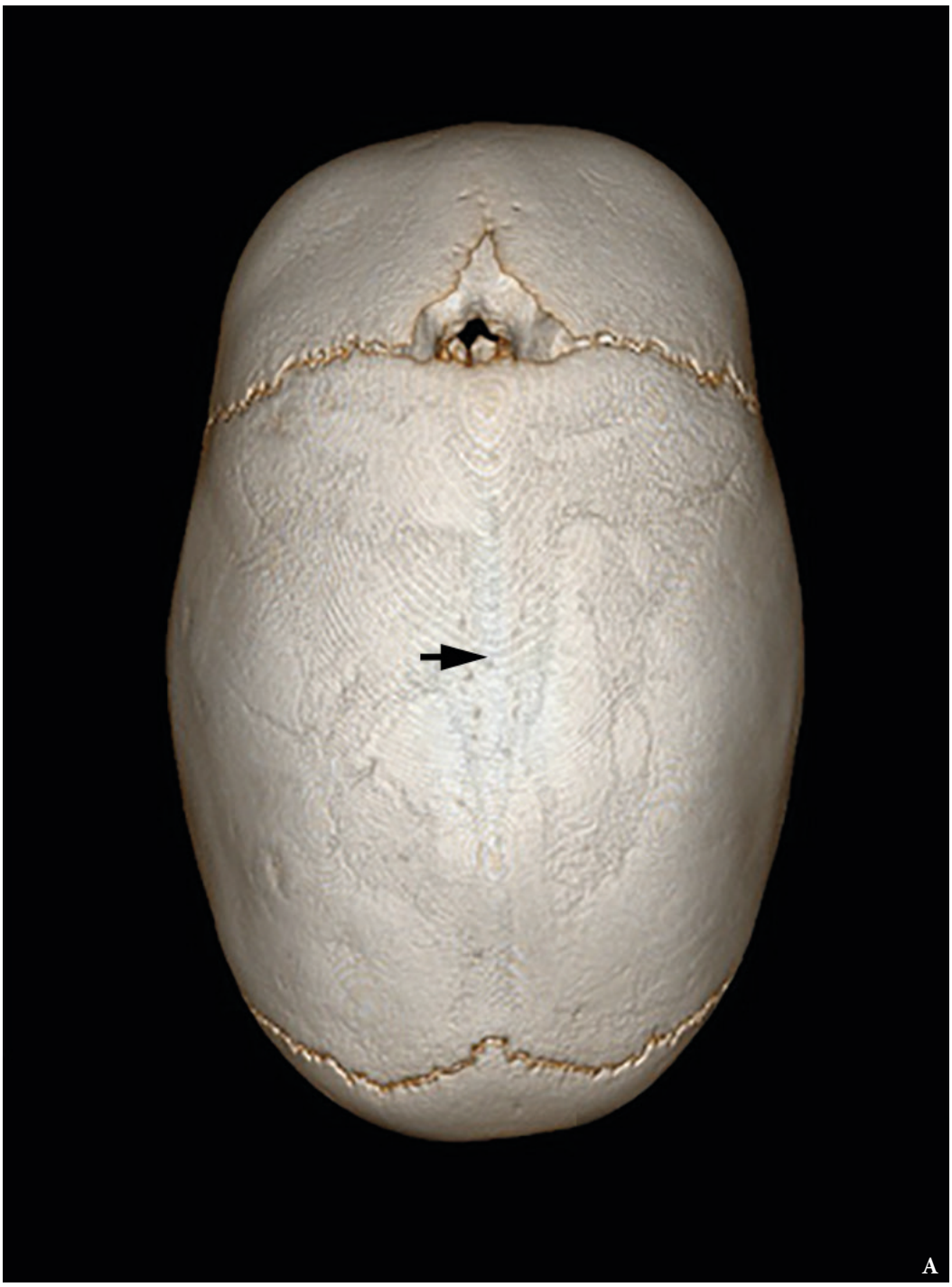

FIGURE 1. Superior view (**A**) of 3D-CT volume-rendered image of a 6-month-old boy with premature, complete fusion (*arrow*) of the sagittal suture. It results in increased anteroposterior diameter, along with bitemporal narrowing due to restriction of biparietal growth. Prominent ridging of the sagittal suture is observed, as well as frontal and occipital bulging. **(Fig 1 continued on next page.)**

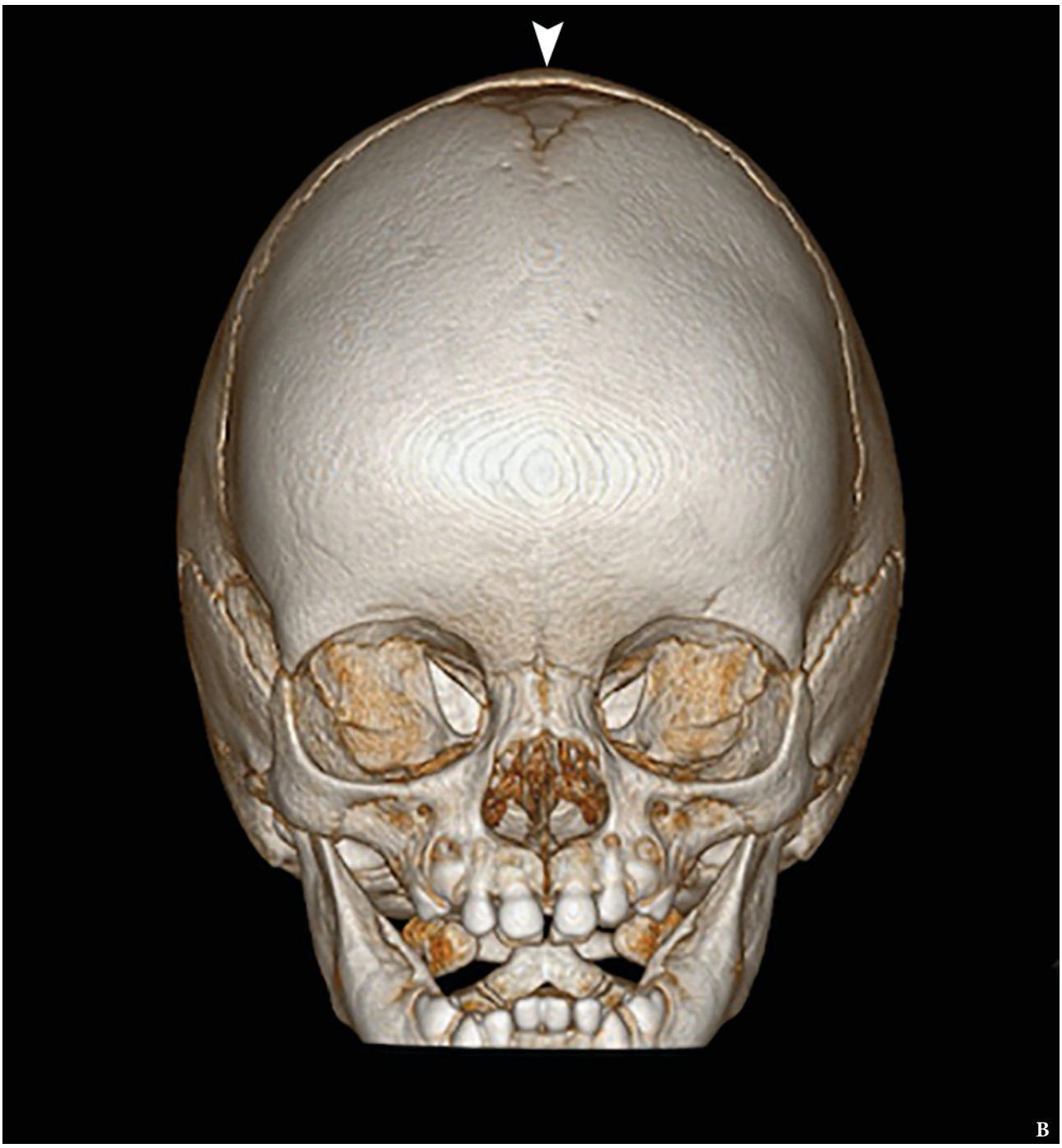

FIGURE 1 (cont'd). Anterior view (**B**) of 3D-CT volume-rendered image of a 6-month-old boy with premature, complete fusion of the sagittal suture. It results in increased anteroposterior diameter, along with bitemporal narrowing due to restriction of biparietal growth. Prominent ridging (*arrowhead*) of the sagittal suture is observed, as well as frontal and occipital bulging. **(Fig 1 continued on next page.)**

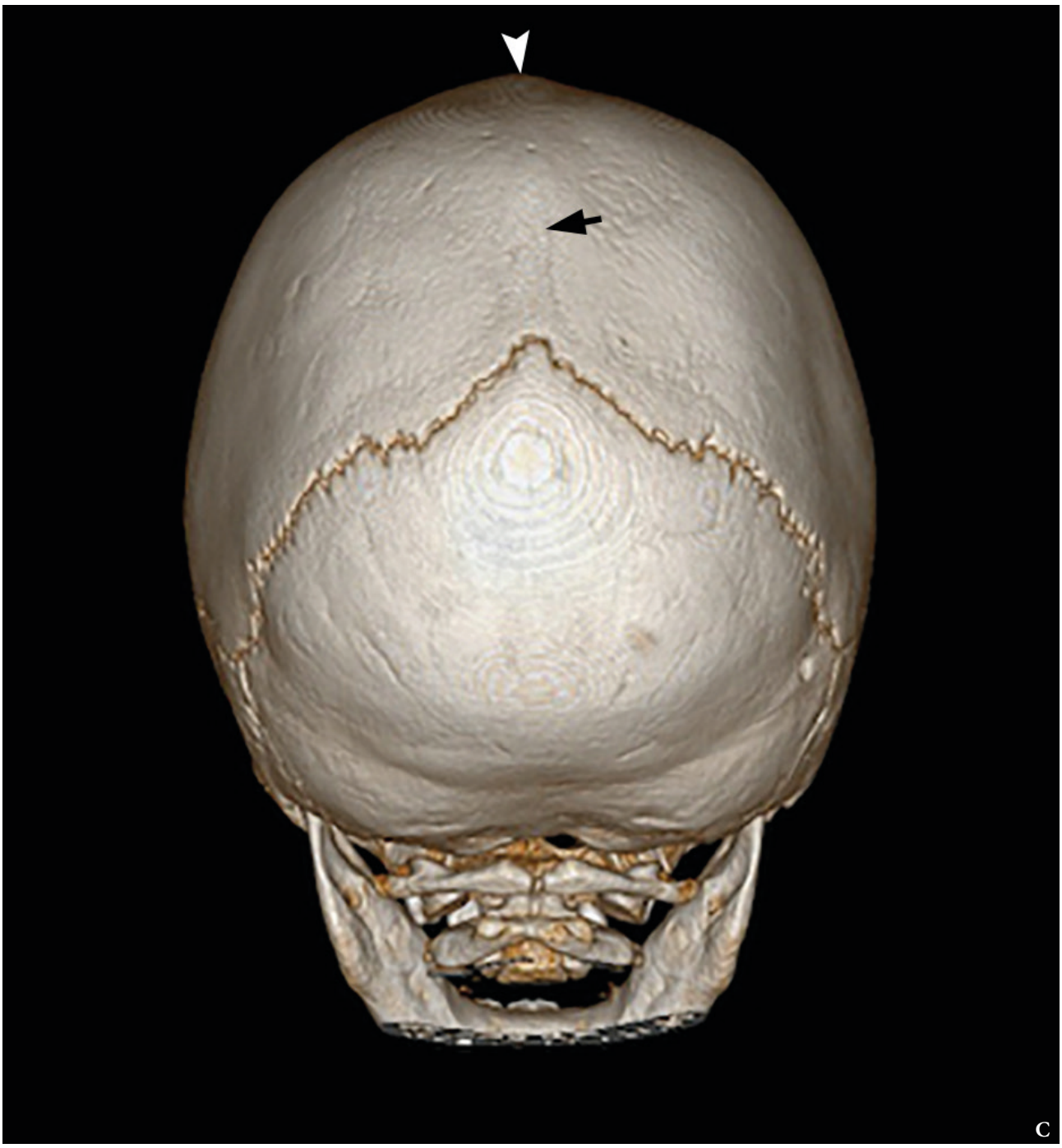

FIGURE 1 (cont'd). Posterior view (**C**) of 3D-CT volume-rendered image of a 6-month-old boy with premature, complete fusion (*arrow*) of the sagittal suture. It results in increased anteroposterior diameter, along with bitemporal narrowing due to restriction of biparietal growth. Prominent ridging (*arrowhead*) of the sagittal suture is observed, as well as frontal and occipital bulging. **(Fig 1 continued on next page.)**

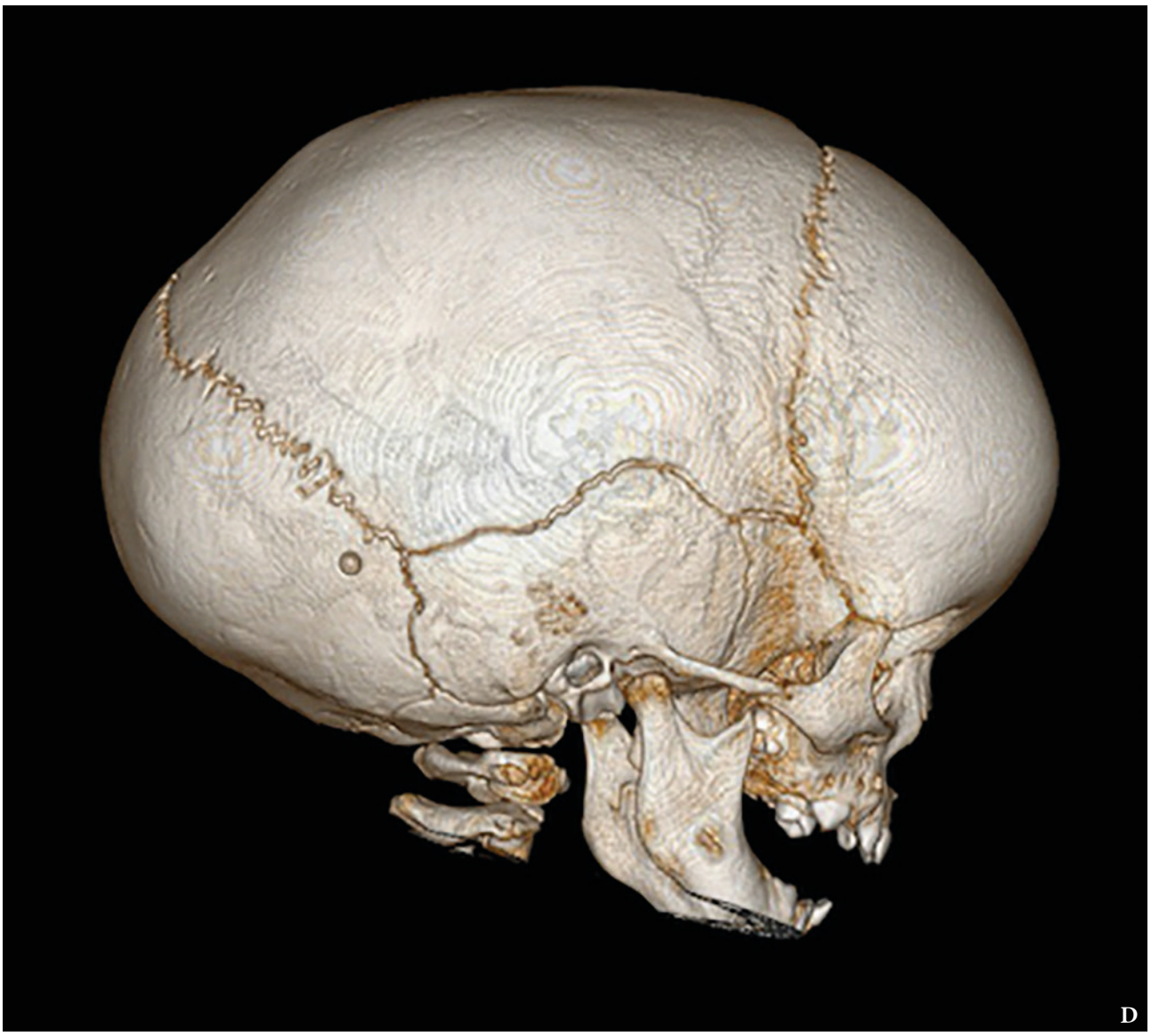

FIGURE 1 (cont'd). Lateral view (**D**) of 3D-CT volume-rendered image of a 6-month-old boy with premature, complete fusion of the sagittal suture. It results in increased anteroposterior diameter, along with bitemporal narrowing due to restriction of biparietal growth. Prominent ridging of the sagittal suture is observed, as well as frontal and occipital bulging.

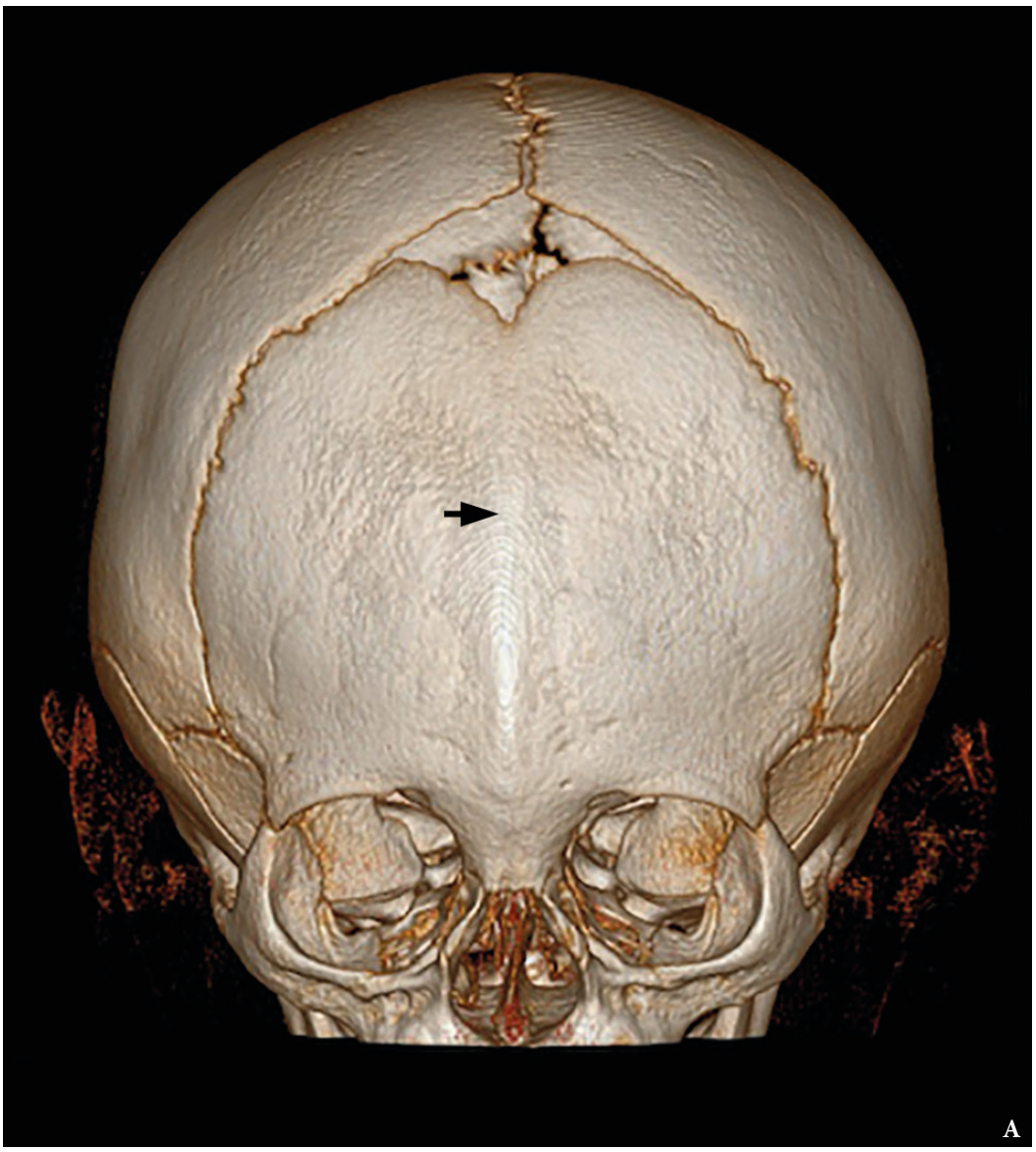

FIGURE 2. Anterior view (**A**) of 3D-CT volume-rendered image of a 4-month-old boy, showing premature fusion of the metopic suture (*arrow)* and the consequent triangular shape of the forehead, together with hypotelorism, flattening of the two frontal bones and bossing of the parieto-occipital regions. A wormian or intrasutural bone is present as well in the lambdoid suture.
(Fig 2 continued on next page.)

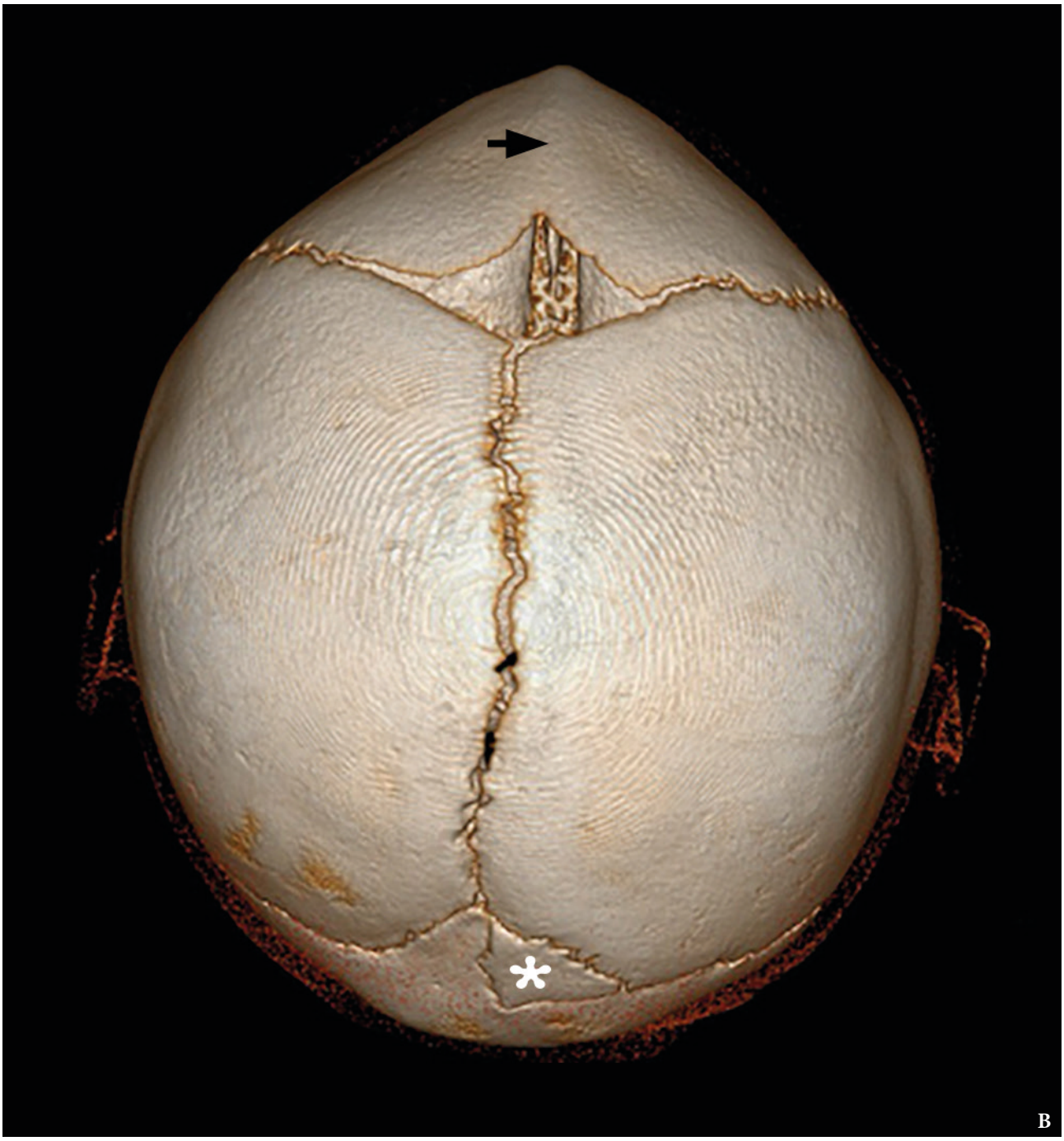

FIGURE 2 (cont'd). Superior view (**B**) of 3D-CT volume-rendered image of a 4-month-old boy, showing premature fusion of the metopic suture and the consequent triangular shape of the forehead, together with hypotelorism, flattening of the two frontal bones and bossing of the parieto-occipital regions. A wormian or intrasutural bone (*asterisk*) is present as well in the lambdoid suture.**(Fig 2 continued on next page.)**

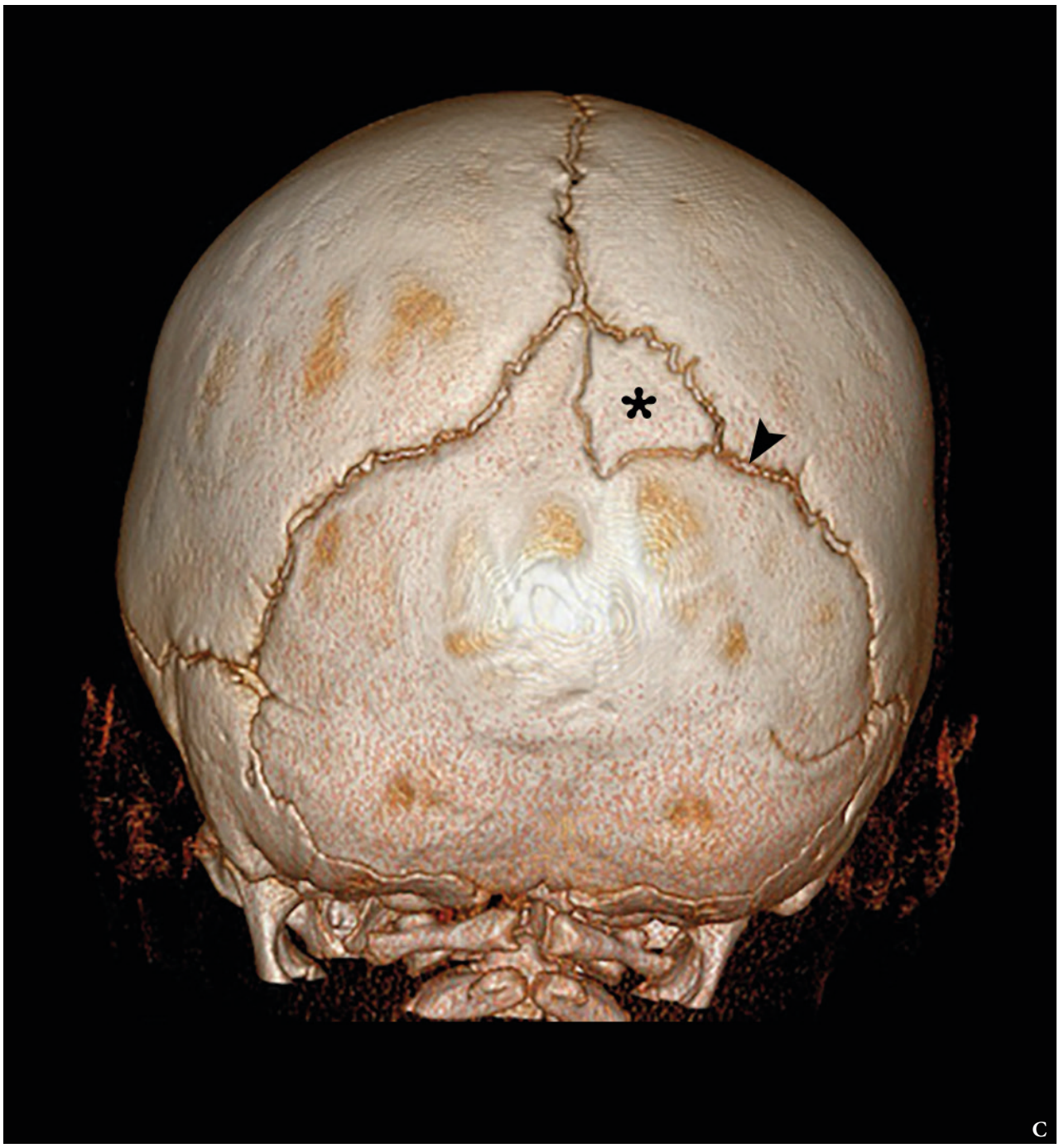

FIGURE 2 (cont'd). Posterior view (**C**) of 3D-CT volume-rendered image of a 4-month-old boy, showing premature fusion of the metopic suture and the consequent triangular shape of the forehead, together with hypotelorism, flattening of the two frontal bones and bossing of the parieto-occipital regions. A wormian or intrasutural bone (*asterisk*) is present as well in the lambdoid suture (*arrowhead*). **(Fig 2 continued on next page.)**

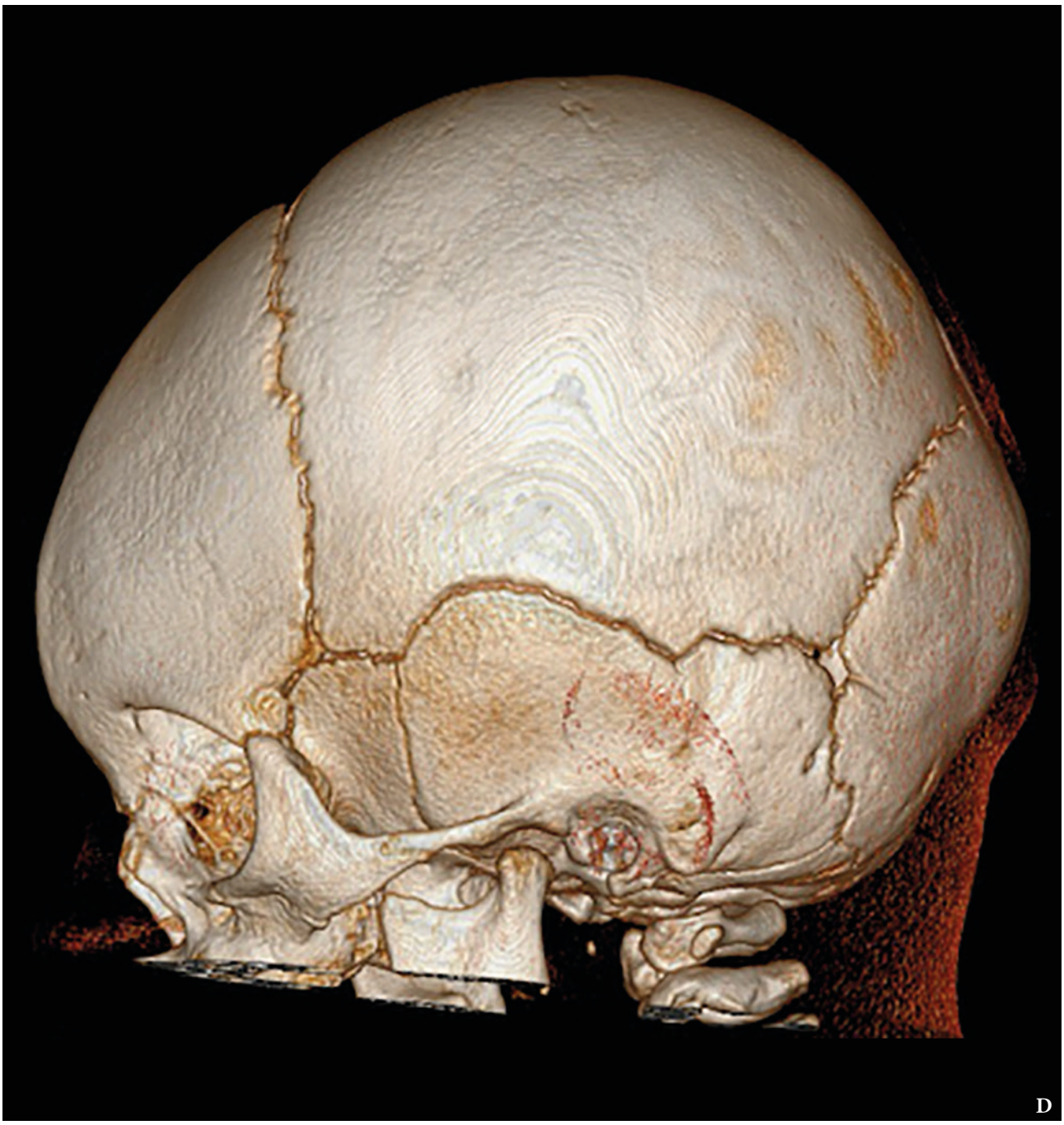

FIGURE 2 (cont'd). Left lateral view (**D**) of 3D-CT volume-rendered image of a 4-month-old boy, showing premature fusion of the metopic suture and the consequent triangular shape of the forehead, together with hypotelorism, flattening of the two frontal bones and bossing of the parieto-occipital regions. A wormian or intrasutural bone is present as well in the lambdoid suture.

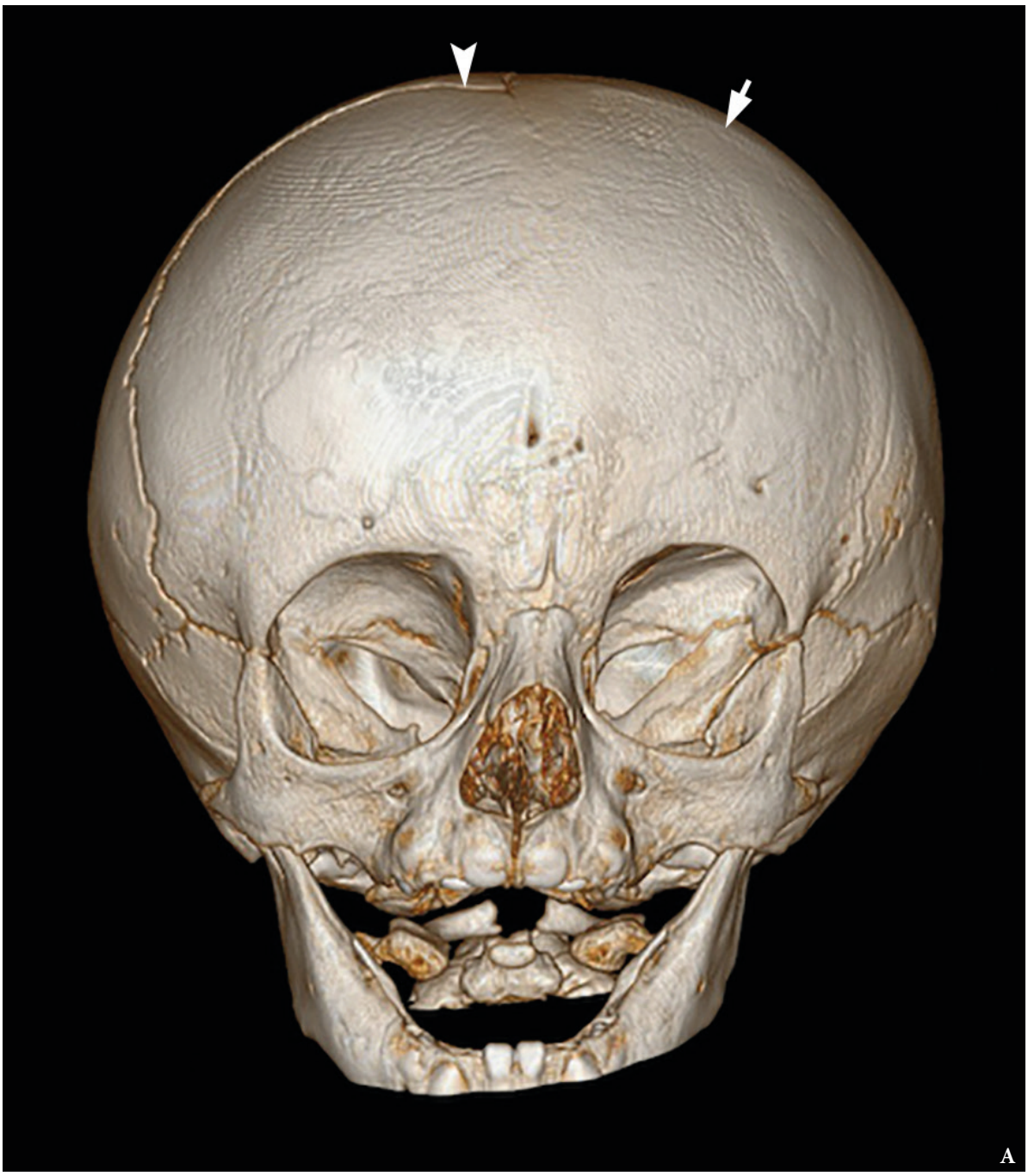

FIGURE 3. Anterior view (**A**) of 3D-CT volume-rendered image of a 7-month-old boy with anterior plagiocephaly as a result of left coronal suture fusion (*arrow*) which leads in flattening of the frontal bone on the affected side, and prominent frontal bossing to the contralateral side. There is also evident orbit asymmetry. The right coronal suture (*arrowhead*) is still open. **(Fig 3 continued on next page.)**

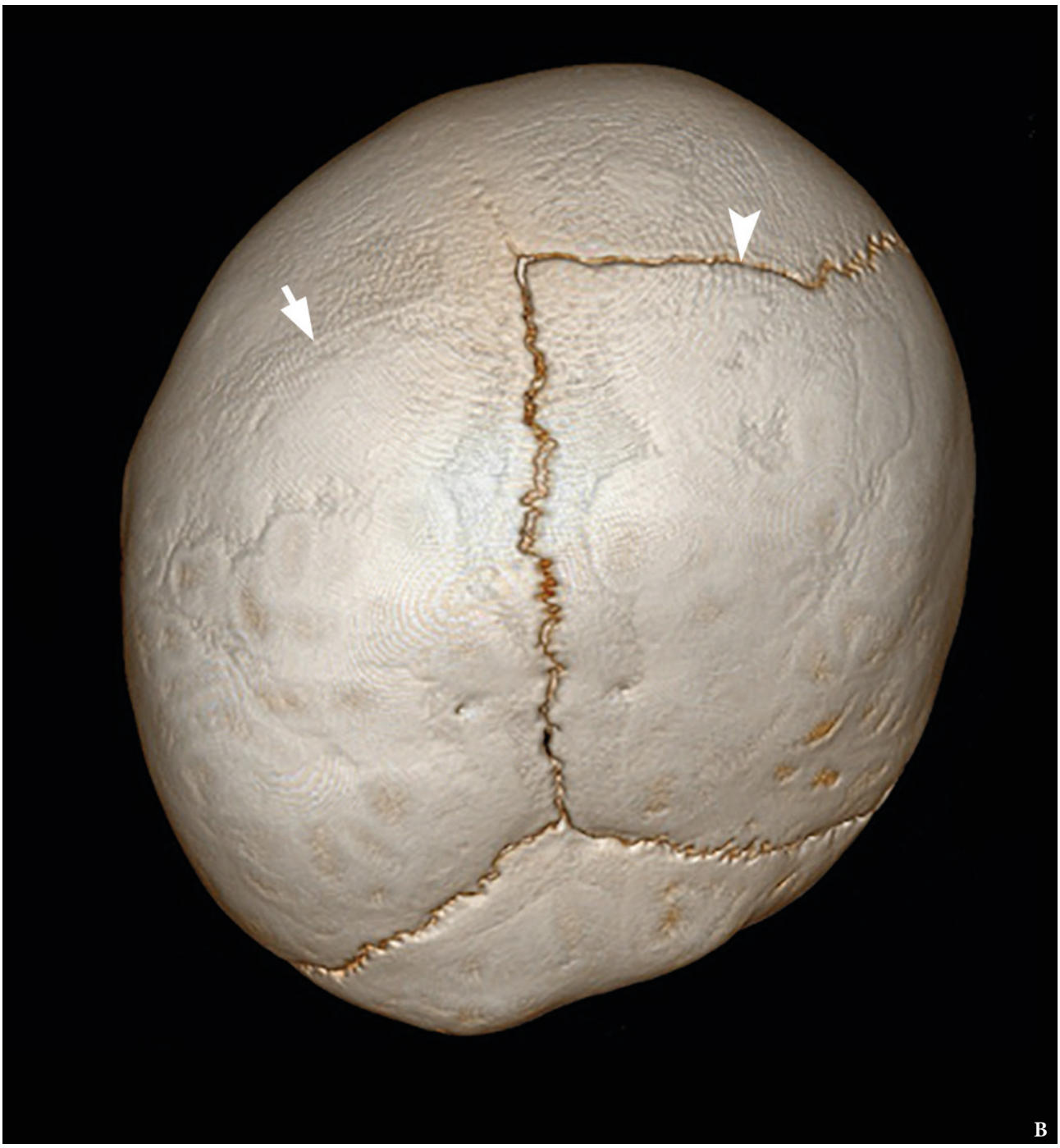

FIGURE 3 (cont'd). Superior view (**B**) of 3D-CT volume-rendered image of a 7-month-old boy with anterior plagiocephaly as a result of left coronal suture fusion (*arrow*) which leads in flattening of the frontal bone on the affected side, and prominent frontal bossing to the contralateral side. There is also evident orbit asymmetry. The right coronal suture is still open (*arrowhead*). **(Fig 3 continued on next page.)**

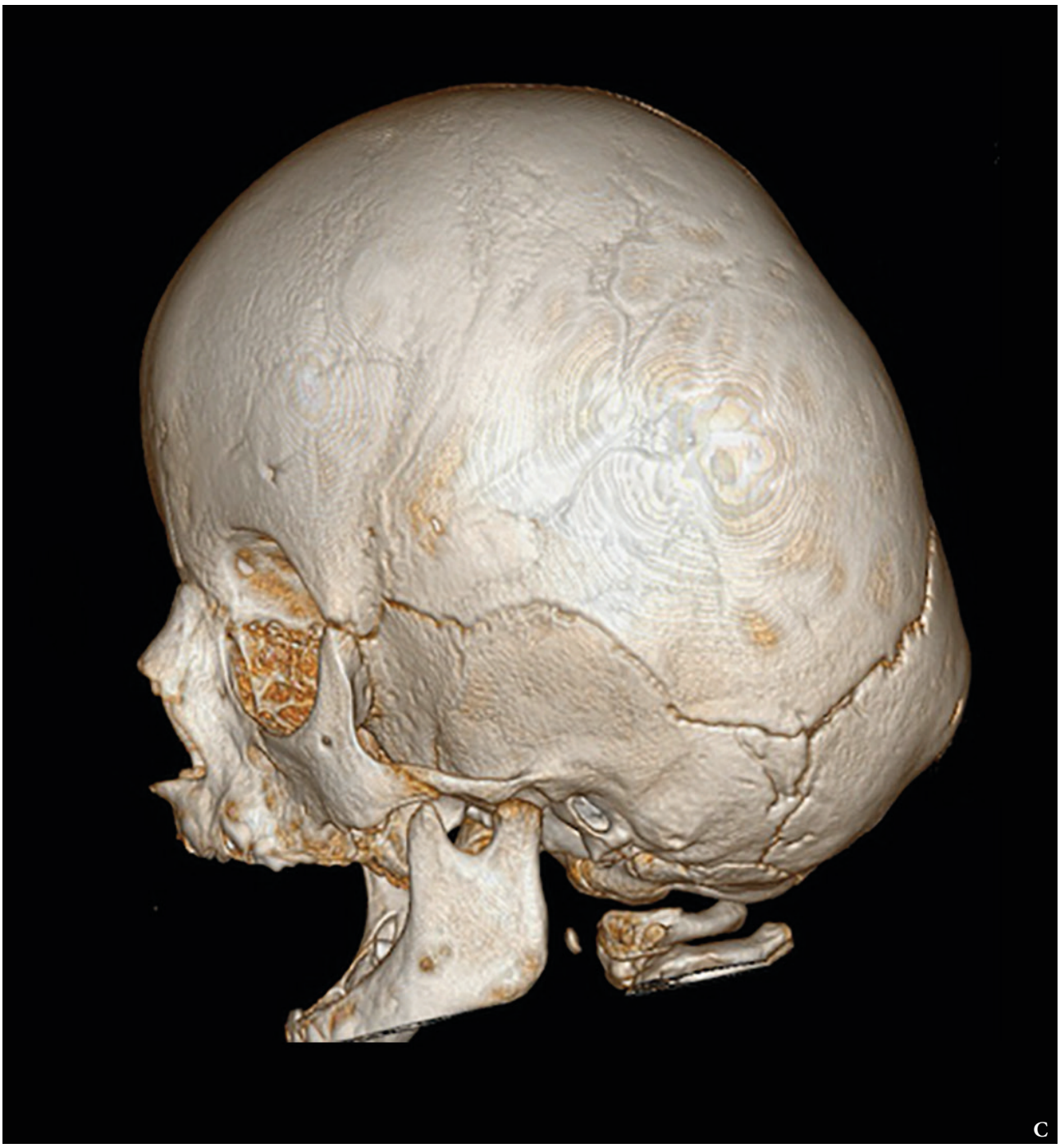

FIGURE 3 (cont'd). Left lateral view (**C**) of 3D-CT volume-rendered image of a 7-month-old boy with anterior plagiocephaly as a result of left coronal suture fusion which leads in flattening of the frontal bone on the affected side, and prominent frontal bossing to the contralateral side. There is also evident orbit asymmetry. The right coronal suture is still open. **(Fig 3 continued on next page.)**

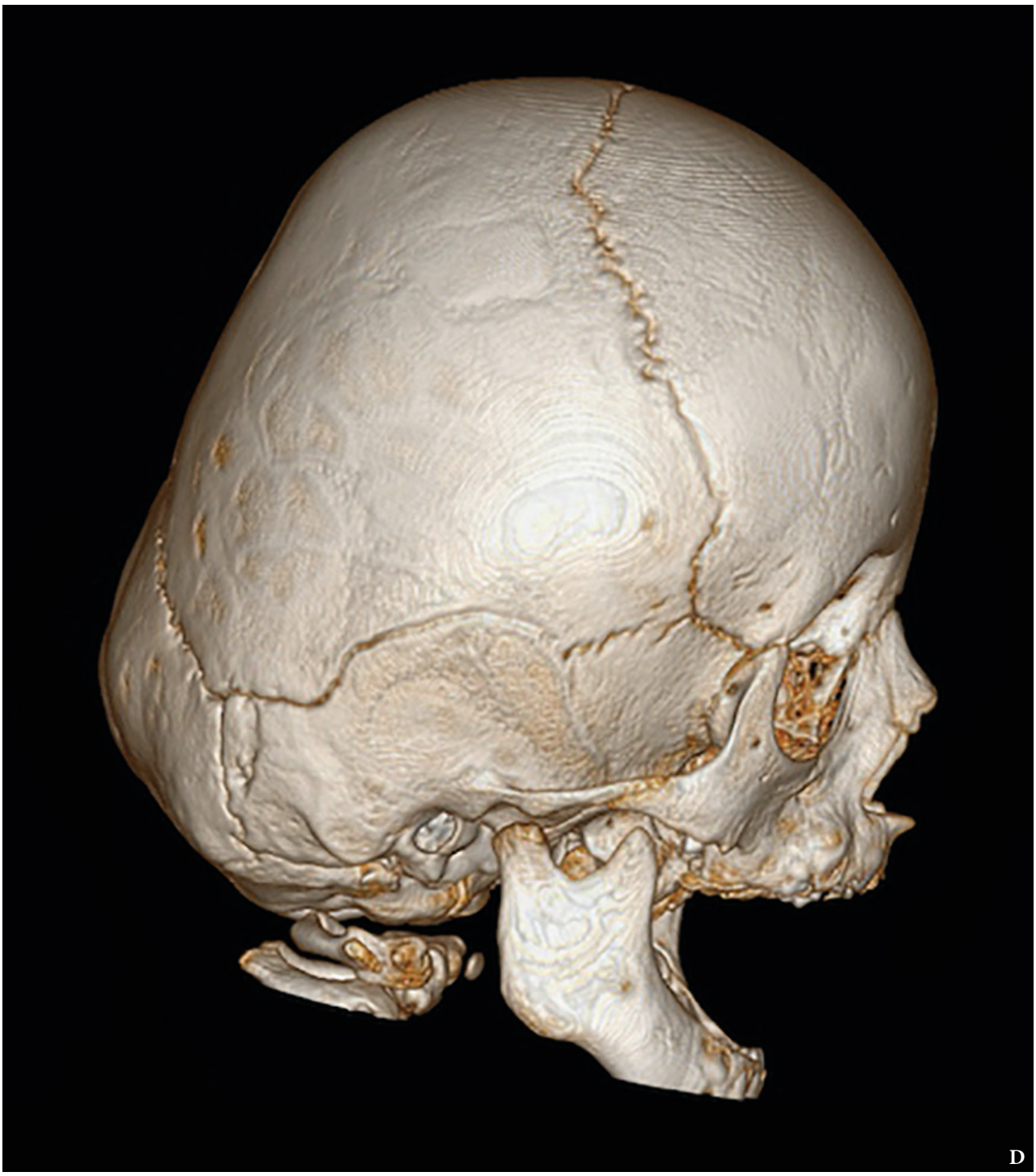

FIGURE 3 (cont'd). Right lateral view (**D**) of 3D-CT volume-rendered image of a 7-month-old boy with anterior plagiocephaly as a result of left coronal suture fusion which leads in flattening of the frontal bone on the affected side, and prominent frontal bossing to the contralateral side. There is also evident orbit asymmetry. The right coronal suture is still open.

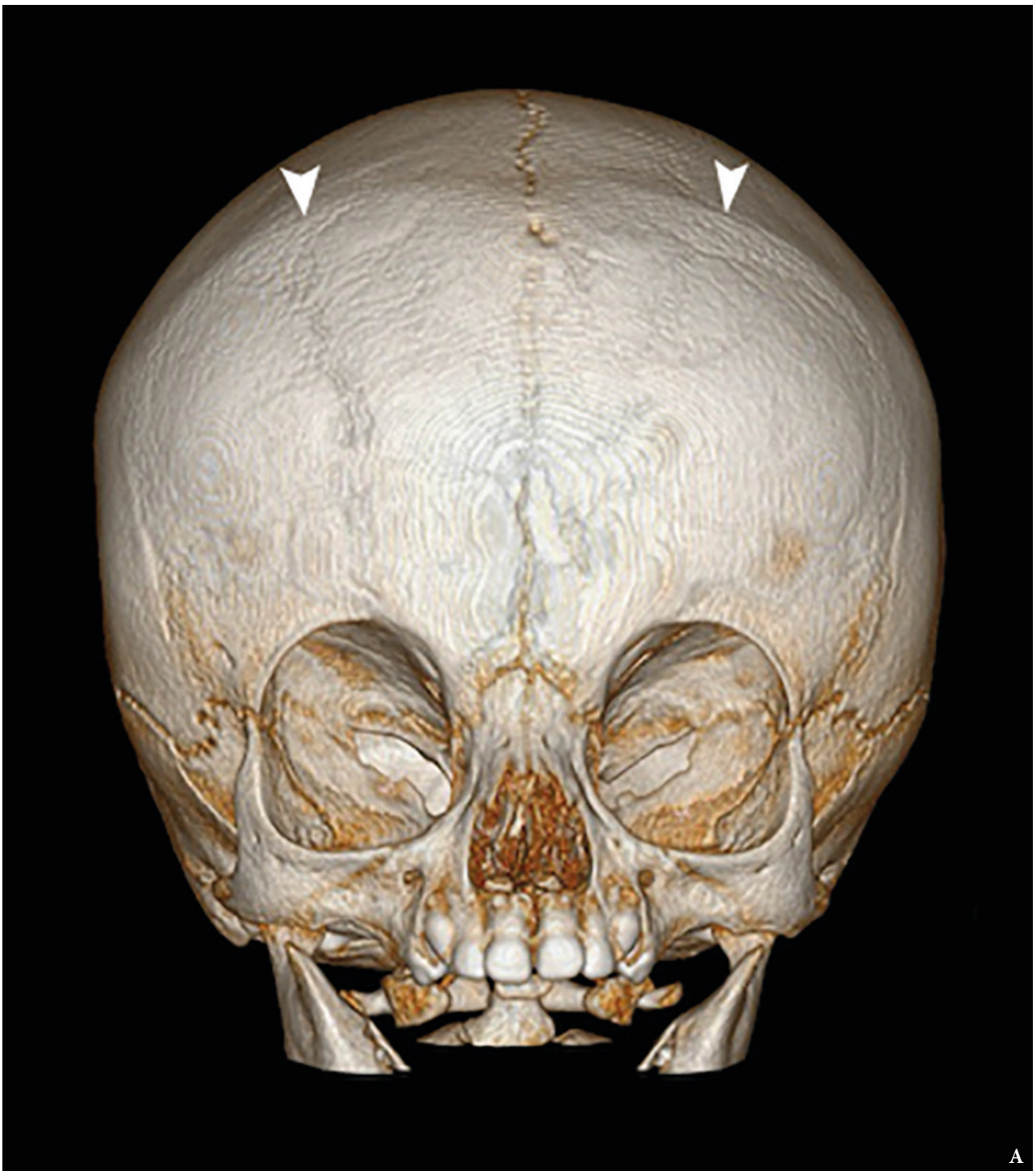

FIGURE 4. Anterior view (**A**) of 3D-CT volume-rendered image of a 6-month-old girl with complete bicoronal suture fusion (*arrowheads*) resulting to restriction of the anteroposterior calvarial growth and pronounced biparietal growth. A consequent prominent frontal bone and an occiput flattening are also evident. **(Fig 4 continued on next page.)**

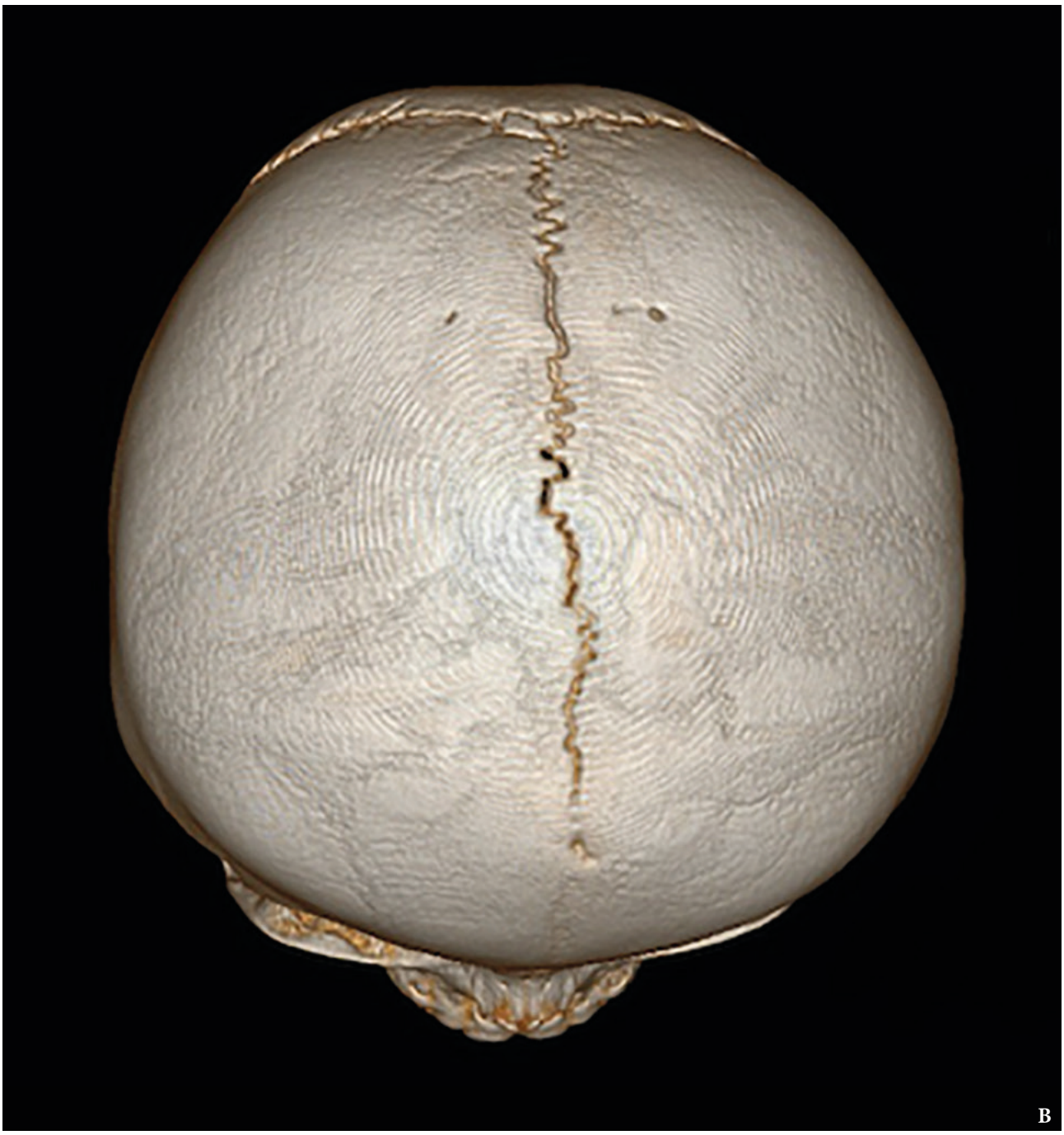

FIGURE 4 (cont'd). Superior view (**B**) of 3D-CT volume-rendered image of a 6-month-old girl with complete bicoronal suture fusion resulting to restriction of the anteroposterior calvarial growth and pronounced biparietal growth. A consequent prominent frontal bone and an occiput flattening are also evident. **(Fig 4 continued on next page.)**

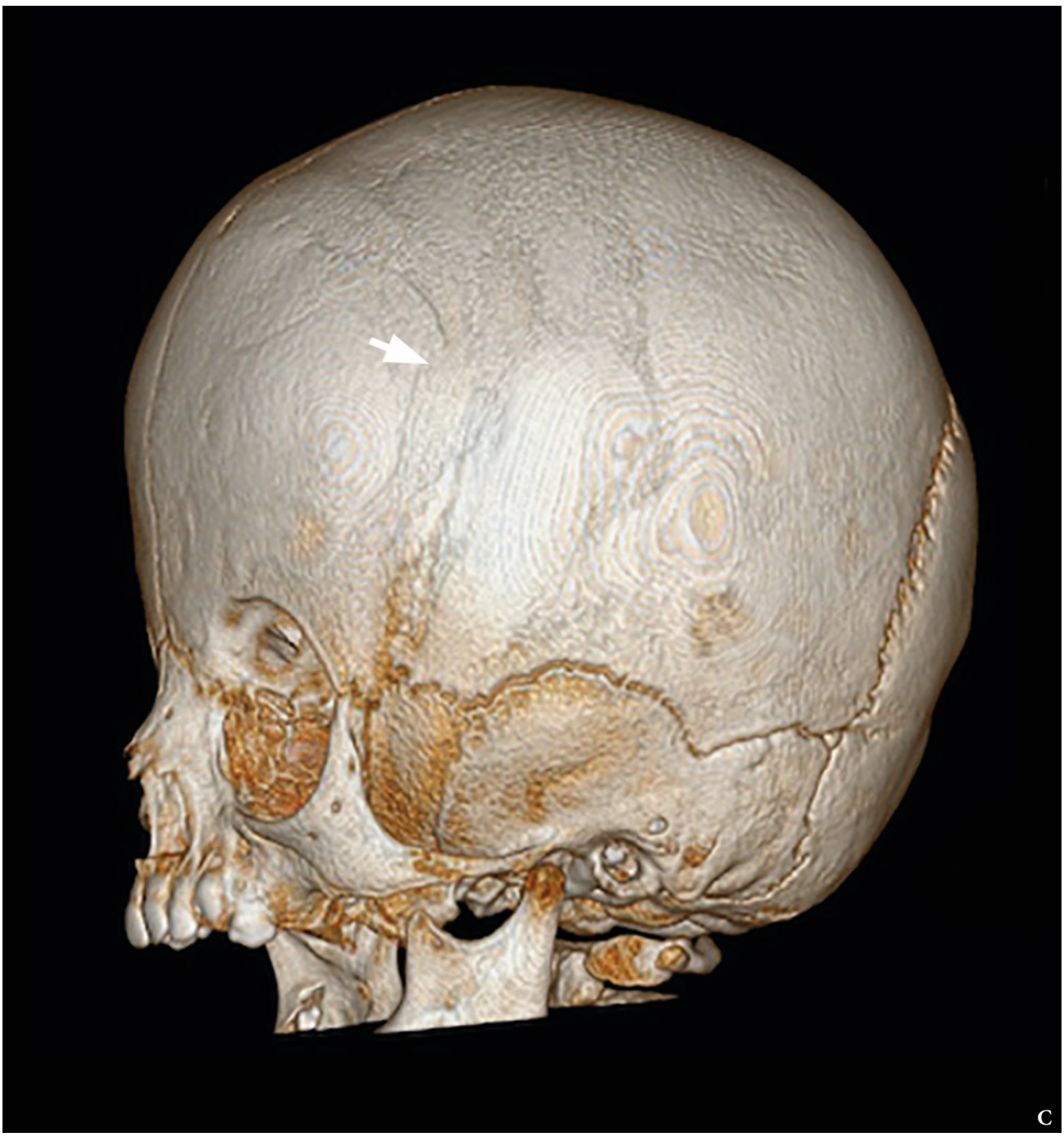

FIGURE 4 (cont'd). Lateral view (**C**) of 3D-CT volume-rendered image of a 6-month-old girl with complete bicoronal suture fusion (*arrow indicated left coronal suture fusion*) resulting to restriction of the anteroposterior calvarial growth and pronounced biparietal growth. A consequent prominent frontal bone and an occiput flattening are also evident. **(Fig 4 continued on next page.)**

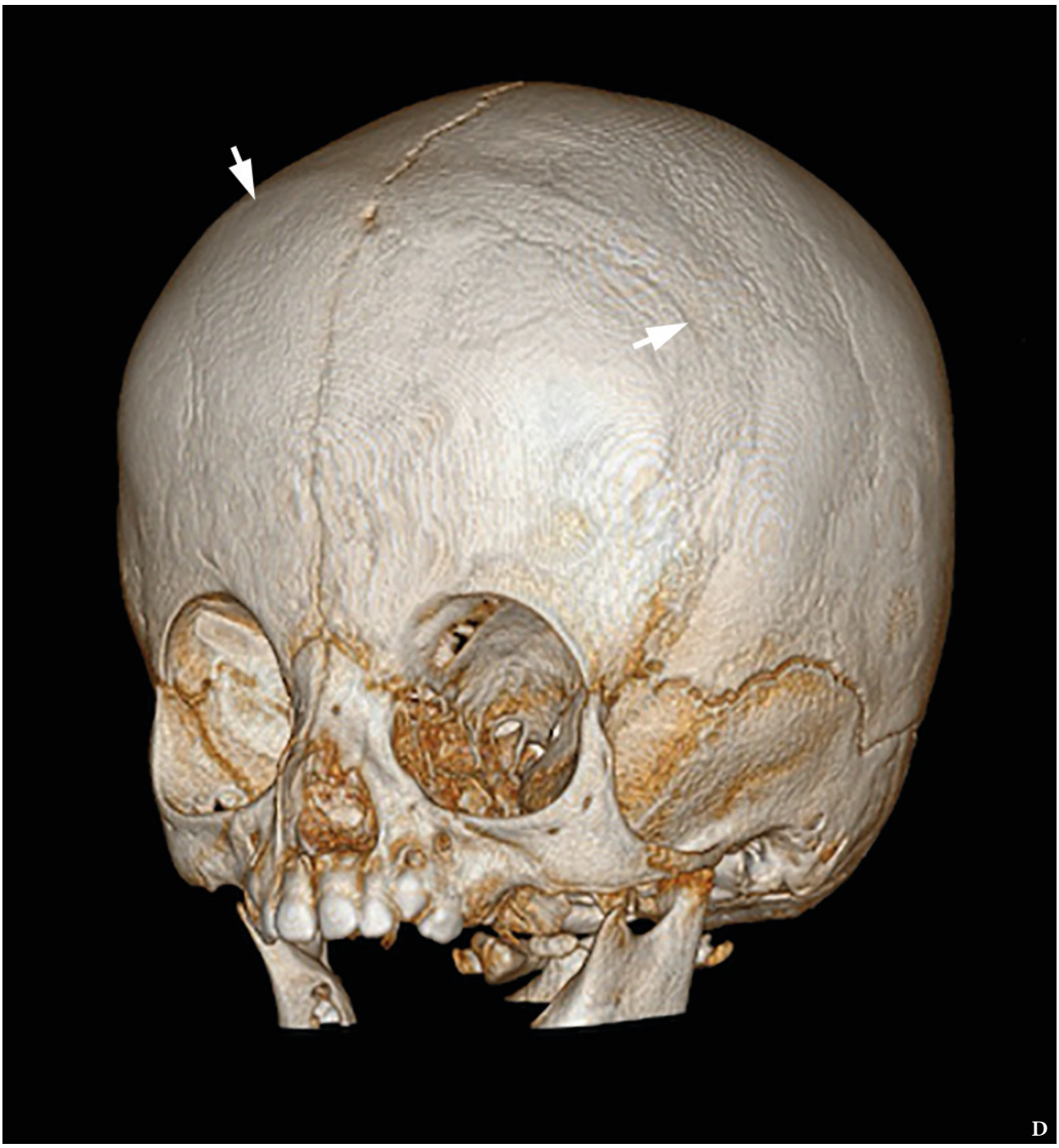

FIGURE 4 (cont'd). Anterolateral view (**D**) of 3D-CT volume-rendered image of a 6-month-old girl with complete bicoronal suture fusion (*arrows*) resulting to restriction of the anteroposterior calvarial growth and pronounced biparietal growth. A consequent prominent frontal bone and an occiput flattening are also evident.

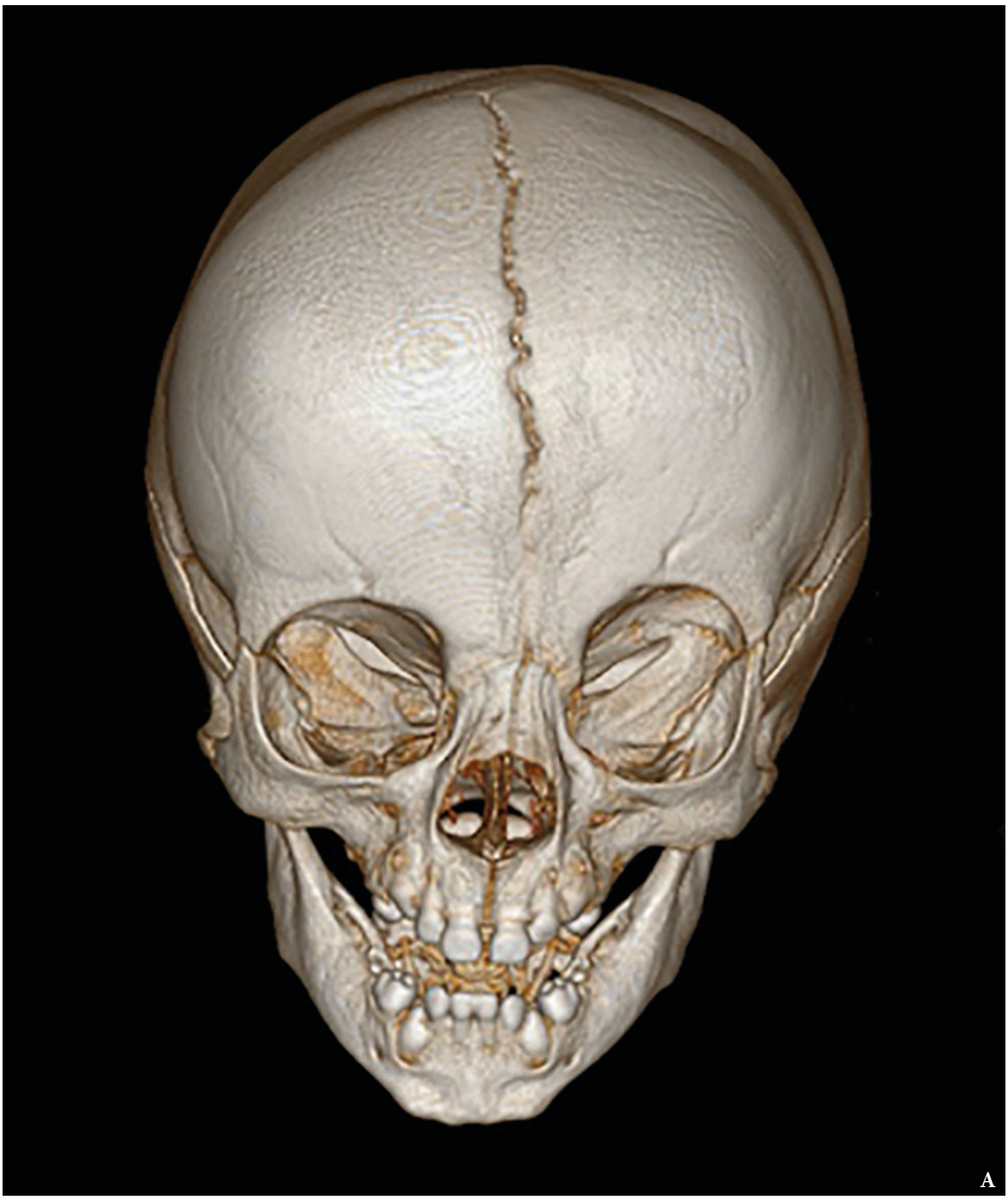

FIGURE 5. Anterior view (**A**) of the 3D-CT volume-rendered image of a 5-month-old girl with premature fusion of the sagittal and left coronal suture ending up with an increased AP dimension, prominent ridging of the sagittal suture, frontal and occipital bulging, flattening of the frontal bone on the affected side and prominent frontal bossing to the contralateral side together with orbit asymmetry. **(Fig 5 continued on next page.)**

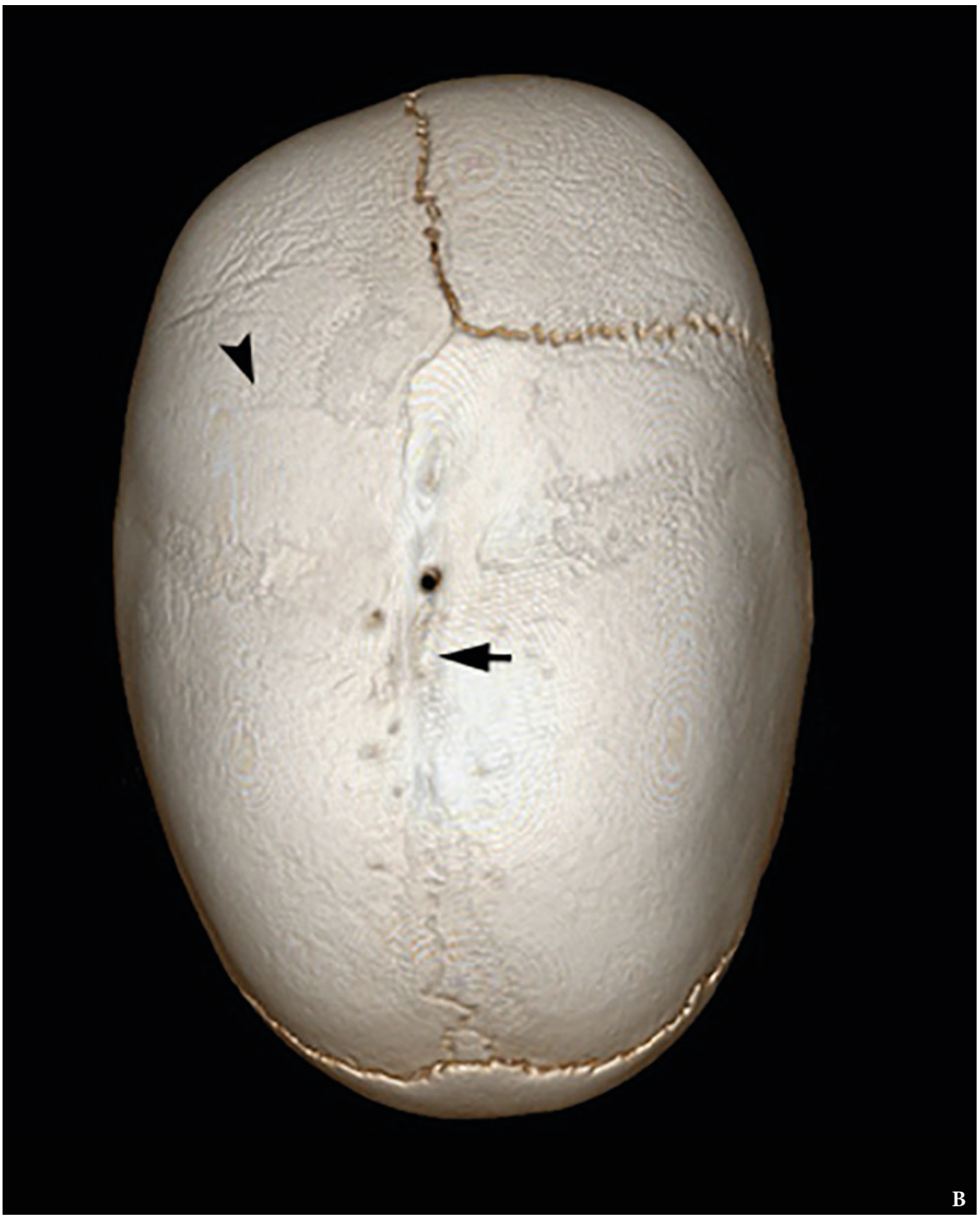

FIGURE 5 (cont'd). Superior view (**B**) of 3D-CT volume-rendered image of a 5-month-old girl with premature fusion of the sagittal (*arrow*) and left coronal (*arrowhead*) suture ending up with an increased AP dimension, prominent ridging of the sagittal suture, frontal and occipital bulging, flattening of the frontal bone on the affected side and prominent frontal bossing to the contralateral side together with orbit asymmetry. **(Fig 5 continued on next page.)**

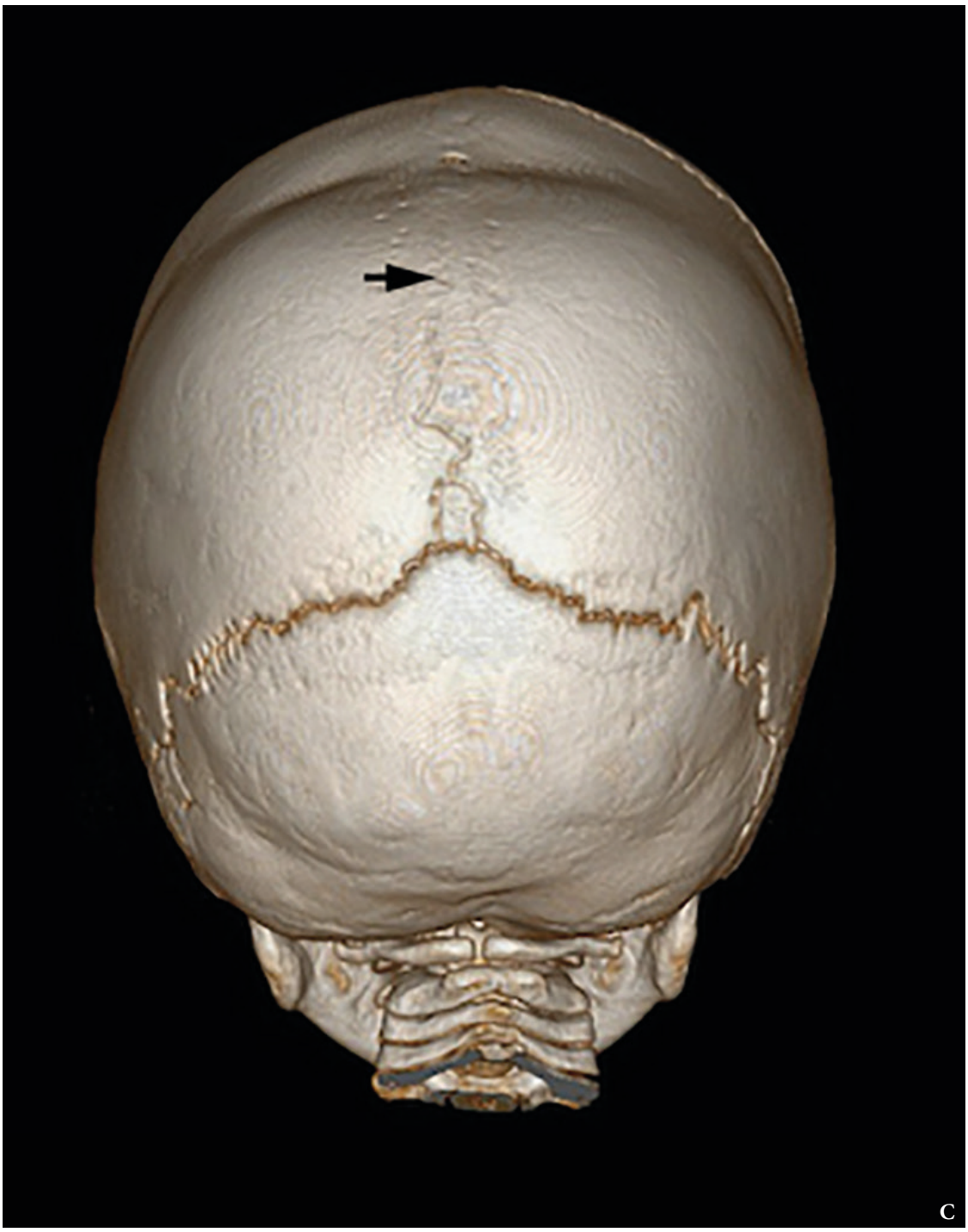

FIGURE 5 (cont'd). Posterior view (**C**) of 3D-CT volume-rendered image of a 5-month-old girl with premature fusion of the sagittal (*arrow*) and left coronal suture ending up with an increased AP dimension, prominent ridging of the sagittal suture, frontal and occipital bulging, flattening of the frontal bone on the affected side and prominent frontal bossing to the contralateral side together with orbit asymmetry. **(Fig 5 continued on next page.)**

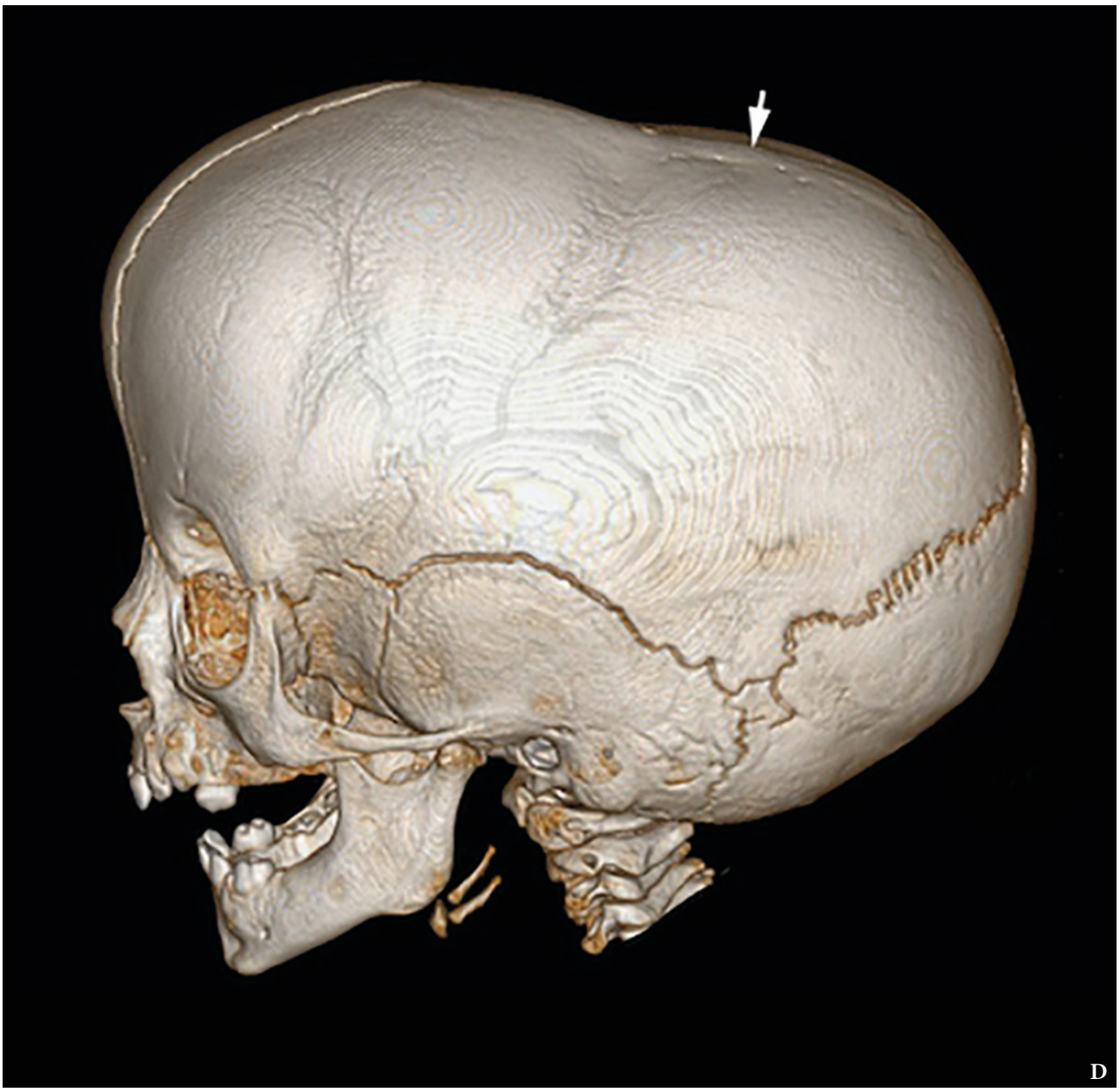

FIGURE 5 (cont'd). Left lateral view (**D**) of 3D-CT volume-rendered image of a 5-month-old girl with premature fusion of the sagittal and left coronal suture ending up with an increased AP dimension, prominent ridging (*arrow*) of the sagittal suture, frontal and occipital bulging, flattening of the frontal bone on the affected side and prominent frontal bossing to the contralateral side together with orbit asymmetry.

Conclusion

Even though craniosynostosis is a well-known entity, at the same time remains a quite demanding disorder as it affects infancy. 3D CT appears as the imaging modality with the best diagnostic performance, but there are still several topics for investigation, as new information is coming up with advancing imaging modalities. The challenges for the diagnosis and treatment of this condition are significant and their consequences will follow the patient throughout the entire life. Thus, every diagnostic intervention has to be cautious and evidence based, in order to have optimal outcomes for the infant. On the domain of diagnosis, there are still questions to be answered, specifically on the possibility to effectively replace diagnostic methods with ionizing radiation (plain radiographs and 3D CT), with other more infant-friendly methods, such as cranial ultrasound and MRI. For this reason large, prospective and multicenter studies are needed in order to reliably establish such diagnostic protocols.

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Conflict of Interests

The authors declare no conflict of interest.

Role of Author and Co-authors

Evangelos G. Kilipiris (concept, design of the paper and writing)
Stefan Pavlik (material collection and writing)
Frantisek Horn (material collection and writing)
Michal Kabat (material collection and writing)

Ethical Approval

Approval was obtained from the Institutional Ethical Committee.

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