

## Three-dimensional CT imaging in pediatric calvarial pathologies

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### ABSTRACT

In children with suspected cranial pathologies, three-dimensional (3D) computed tomography (CT) imaging is superior to other modalities. It can help differentiate actual pathology from normal or variant appearances. Sutures and fontanelles, synostosis, abnormalities of head shape without craniosynostosis, congenital calvarial defects, cranial fractures, bone tumors, and postoperative cranial vault can be assessed easily with 3D CT imaging. We aimed to discuss the common normal, variant, and pathological findings that 3D CT imaging can aid to diagnose as well as explain the usefulness of 3D CT imaging in the diagnosis of calvarial pathologies.

In children with suspected cranial pathologies, three-dimensional (3D) computed tomography (CT) is superior to other modalities. Three-dimensional CT can help differentiate actual pathology from normal or variant appearances. Sutures and fontanelles, synostosis, abnormalities of head shape without craniosynostosis, congenital calvarial defects, cranial fractures, bone tumors, and postoperative cranial vault can be assessed easily using 3D CT.

We aimed to discuss the common normal, variant, and pathological findings that 3D CT can assist diagnosis of, as well as explain the usefulness of 3D CT in the diagnosis of calvarial pathologies.

### Multidetector CT scanning technique

All CT examinations were performed using a 64-slice CT scanner (Aquillon 64, Toshiba Medical Systems, Tochigi, Japan). The scanning parameters included 120 kV, 100–120 mA, section thickness of 0.5 mm, and reconstruction interval of 0.5 mm. The scan revolution time was 0.5 s. Three-dimensional reconstructions were generated on the CT scanner console and sent to a picture archiving and communication system. For patients who required detailed evaluation, 3D volume-rendered (VR) and 3D maximum intensity projection (MIP) images were evaluated in a workstation (Aquarius workstation, TeraRecon, San Mateo, California, USA).

### Embryology and anatomy

The development of the skull is outlined in Fig. 1. Calvaria is a Latin term that refers to the upper part of the head that surrounds the brain and special sense organs. It is formed by pressure of the growing cerebral and cerebellar hemispheres with the dura playing a regulatory role in this process (1).

Membranous bones of the vault are separated by sutures that facilitate vaginal passage and allow uniform growth of the calvarium by its fibrous connective tissue content. The growth of the skull is perpendicular to the suture lines and parallel to a fused suture (Virchow's law). If there is premature fusion of a suture, the calvaria show no growth perpendicular to the affected suture (1).

The anterior fontanelle is a space in the intersection of the sagittal, coronal, and metopic sutures and closes typically by 12 months of age. The posterior fontanelle is in the conjunction of the sag-

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ittal and lambdoid sutures and closes by about three months of age (1–3). The closure of the sutures and fontanelles are outlined in Table. Fig. 2 shows the normal calvarial 3D anatomy.

### Wormian bones (intrasutural bones)

Wormian bones are accessory bones that occur within the cranial suture and fontanelles, most commonly within the posterior sutures (Fig. 3).

Wormian bones are usually considered as normal variants, but sometimes they are associated with cleidocranial dysplasia, pycnodysostosis, osteogenesis imperfecta, hypothyroidism, hypophosphatasia, acro-osteolysis, and Down's syndrome (4).

A larger, single, centrally located intrasutural bone at the junction of the lambdoid and sagittal suture is called *os incae* (interparietal bone) (Fig. 4). It is formed in a persistent mendosal suture (5).

### Lacunar skull, increased convolutional markings, and copper beaten skull

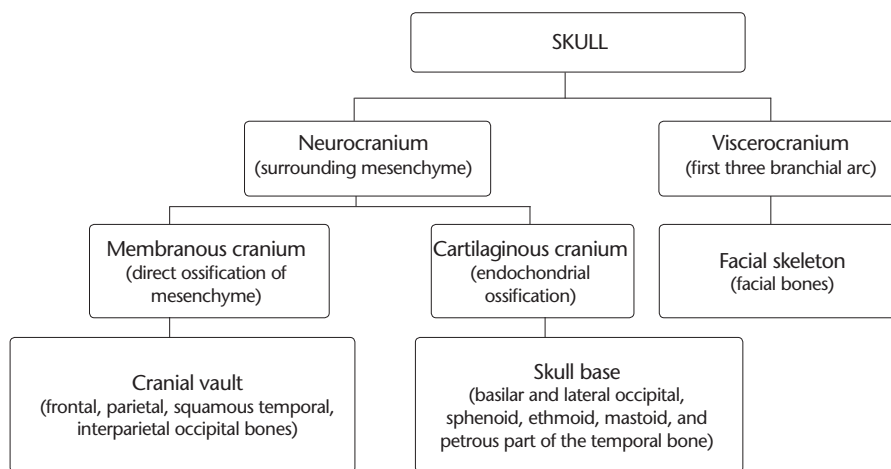
Lacunar skull, increased convolutional markings, and copper beaten skull are confusing terms.

Lacunar skull is a dysplasia of the membranous bone. The well-defined lucent areas in the calvarium represent nonossified fibrous bones, which are bound by normally ossified bones (2). They are usually present at birth and occur most prominently in the parietal and occipital bones. The inner table is more affected than the outer table. The lacunae resolves spontaneously by the age of six months and is not related to the degree of hydrocephalus. Lacunar skull is usually associated with Chiari II malformation and less commonly with encephalocele (2).

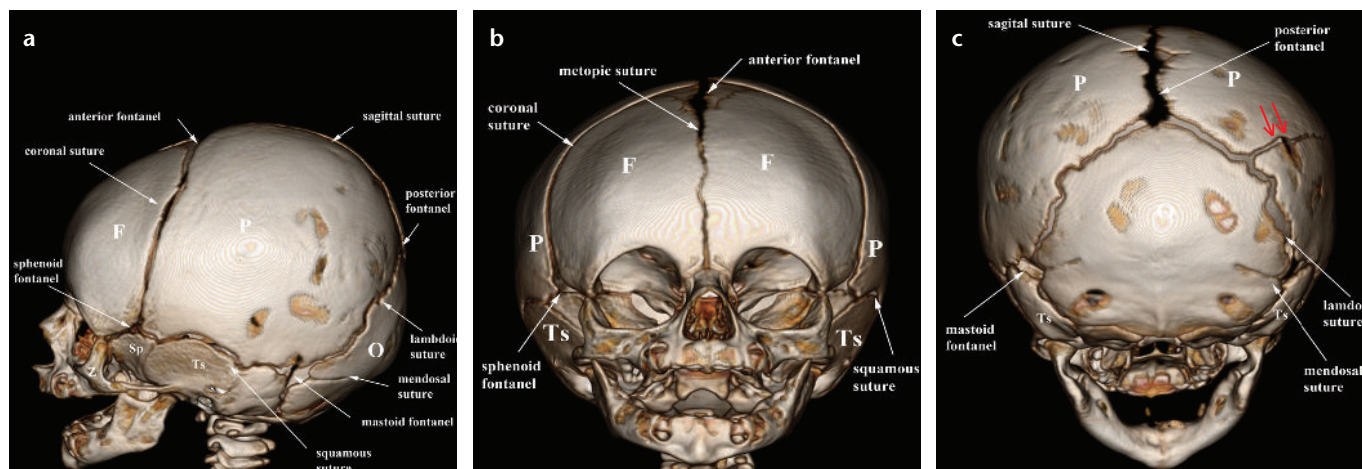
Convolutional markings are inner table indentations that are caused

**Table.** Normal age of the fontanelle/suture closure

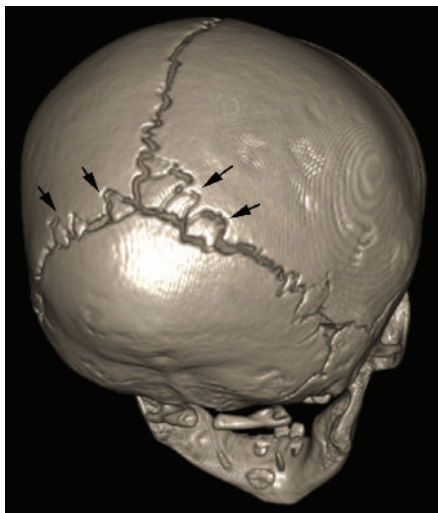
Fontanelle/suture	Age of the closure
Anterior fontanelle	15–18 months
Posterior fontanelle	3–6 months
Posterolateral fontanelle (mastoid)	2 years
Anterolateral fontanelle (sphenoidal)	3 months
Metopic suture	9–11 months (may persist into adulthood)
Sagittal suture	30–40 years
Coronal sutures	30–40 years
Lambdoid sutures	30–40 years
Squamosal sutures	30–40 years



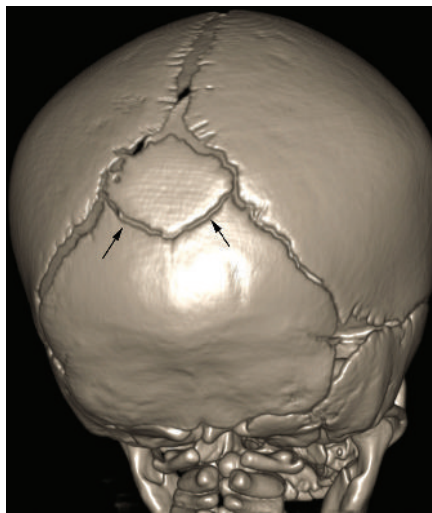
**Figure 1.** Development of the skull.



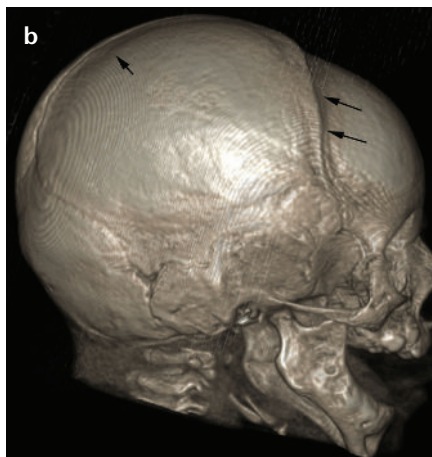
**Figure 2.** a–c. Normal 3D calvarial anatomy from the lateral (a), front (b), and back (c) views. There is a linear fracture in the right parietal bone (c, double arrows). F, frontal bone; O, occipital bone; P, parietal bone; Sp, sphenoid bone; Ts, temporal bone squamous portion.



**Figure 3.** Wormian bones. There are many accessory bones within the lambdoid suture and posterior fontanelle (arrows).



**Figure 4.** *Os incae* (interparietal bone). A large, single intrasutural bone at the junction of the lambdoid suture and sagittal suture (arrows) is seen.



**Figure 5.** a, b. Microcephaly. Axial multiplanar reconstruction (a) and 3D image (b) show that the sutures are closed and overlapping (b, arrows) in a one-year-old patient due to severe hypoxic-ischemic injury.

by the cerebral surface of the growing brain in infants. They occur later than a lacunar skull, particularly during periods of rapid brain growth, between ages 2–3 and 5–7 years. They become less prominent after eight years of age. Convolutional markings are now considered to reflect normal brain growth. If they become prominent and are evident throughout the skull rather than the posterior parts, they reflect a pathologic condition, the so called copper beaten skull (6).

Copper beaten skull is an indicator of chronic elevated intracranial pressure resulting from craniosynostosis, hydrocephalus, and intracranial masses. Macrocrania, splitting of the

sutures, skull demineralization, and erosion or enlargement of the sella turcica may be observed due to increased intracranial pressure (6, 7).

#### Abnormalities in head size (macrocephaly and microcephaly)

Macrocephaly is a disorder characterized by a head larger than two standard deviations from the normal distribution. There are three major causes of macrocephaly: hydrocephalus (increased cerebrospinal fluid), megalencephaly (enlargement of the brain due to neurocutaneous syndromes or metabolic diseases) or thickening of the skull (anemia, rickets, hyperphosphatemia, osteopetrosis, osteogenesis imperfecta, and cleidocranial dysosto-

sis). It may be constitutional or due to benign causes such as benign enlargement of the subarachnoid space. CT is superior to skull radiography because the former can differentiate these major categories (8).

Microcephaly is a condition characterized by a head less than two standard deviations from the normal distribution. The head size is smaller in some ethnic groups, and the condition can be familial. However, it is important to diagnose microcephaly and identify the cause. There are two major causes of microcephaly: primary causes (chromosomal disorders, neurulation defects such as anencephaly and encephalocele, prosencephalization defects such as agenesis corpus callosum and holoprosencephaly, migration defects) and secondary causes (intrauterine infection, toxins and vascular occlusions, severe hypoxic-ischemic injury, and postnatal systemic diseases) (2, 8). Due to the lack of brain growth, the force keeping the cranial bones separated does not exist, and there may be early closure of the sutures or even overlapping of the skull bones (Fig. 5).

#### Abnormalities in head shape

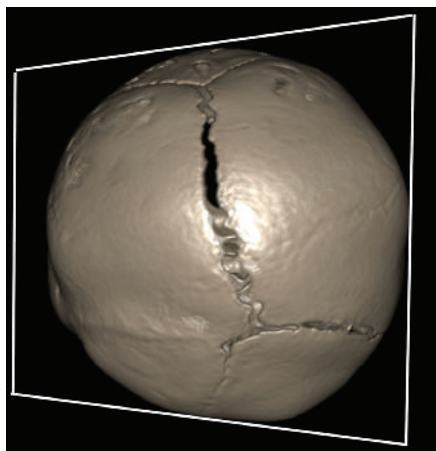
There may be abnormalities in the shape of the neonatal calvaria due to pressure on the head during childbirth. This is called fetal or newborn molding and usually disappears after a few days. Faulty fetal packing indicates concave depressions in the calvaria due to extrinsic pressure of the limb or uterine leiomyoma (2).

Plagiocephaly without craniosynostosis (posterior deformational, positional plagiocephaly) is associated with sleeping position (sleeping on back), congenital torticollis, abnormal vertebra and neurologic deficits. There is ipsilateral frontal and contralateral occipital bossing (parallelogram shape) and anterior displacement of the ipsilateral ear (9). Additionally, there is no significant distortion of the anterior-posterior axis of the skull base (Fig. 6) (10).

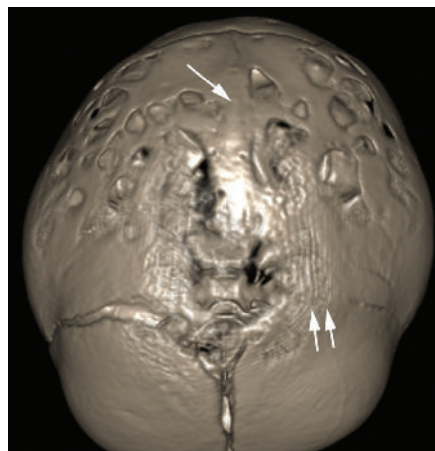




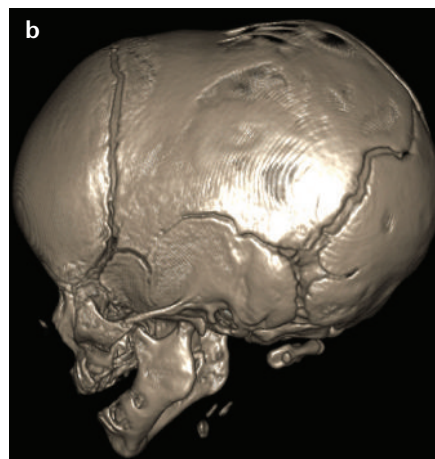
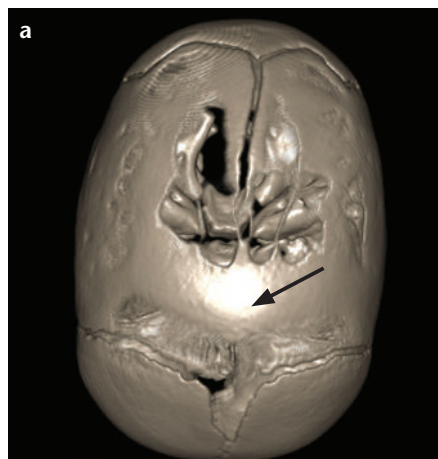
**Figure 6. a–c.** Positional plagiocephaly. The vertex view (a) shows the parallelogram shape of the posterior calvaria with ipsilateral frontal bossing (arrow) and contralateral occipital bossing (double arrows). The skull base view (b) in another patient shows a minimum shift in the midline lines (white, posterior fossa axis line; black, anterior fossa axis line). The long axis of the left temporal bone (black lines) is more anterior than that of the right temporal bone, clinically reflecting anterior displacement of the left ear. There is flattening of the posterior calvaria (b, c, white lines), ipsilateral frontal bossing (b, c, white arrows), contralateral occipital bossing (b, c, double white arrows), and parietal bone fracture parallel to the plane of the imaging (c, black arrows) in the same patient. Convolutional markings are observed as lucent areas in 3D volume-rendered images in the parietal and occipital bones.



**Figure 7.** Plagiocephaly and trapezoid shape on vertex view due to single suture synostosis.



**Figure 8.** Plagiocephaly and copper beaten skull. Premature fusion of the sagittal suture (arrow) and left coronal suture (double arrows). There are multiple lucencies in the parietal and occipital bones due to increased intracranial pressure, copper beaten skull.



**Figure 9. a, b.** Scaphocephaly. Premature fusion of the anterior part of the sagittal suture (arrow) and increased anteroposterior diameter of the skull. There are lucencies in the parietal and occipital bones due to increased intracranial pressure.

### Craniosynostosis

Premature fusion of the sutures is commonly isolated and sporadic (non-syndromic). Craniosynostosis may be associated with some syndromes, including Crouzon, Apert, Pfeiffer, and Carpenter syndromes (1, 11).

Plagiocephaly refers to a skewed or oblique head (1). Unilateral coronal synostosis (anterior plagiocephaly) (Fig. 7), unilateral synostosis of the lambdoid suture (posterior plagiocephaly) or asynchronous synostoses of multiple sutures (Fig. 8). A radiologist should distinguish posterior plagiocephaly, which requires surgery, from positional plagiocephaly, which can be treated conservatively (10, 11). Some important indicators of plagiocephaly include the following:

1) Lambdoid suture synostosis; 3D VR images are useful for rapid assessment of premature fusion. Three-dimensional MIP images, which can be performed within seconds using computer workstations, can be added for further detailed evaluation (12).

2) Contralateral frontal and parietal bossing (trapezoidal shape) and posterior displacement of the ipsilateral ear (9). In positional plagiocephaly, there is ipsilateral frontal and contralateral occipital bossing with

a parallelogram shape and anterior displacement of the ipsilateral ear.

3) In the skull base view, the posterior fossa axis line (central line from the basion to opisthion) will be away from the anterior fossa axis line (central line bisecting the cribriform plate) toward the site of the lambdoid fusion (9, 10). In positional plagiocephaly, the lines are continuous with each other or have minimal deviation ( $2.3^\circ \pm 1.3^\circ$ ) (10).

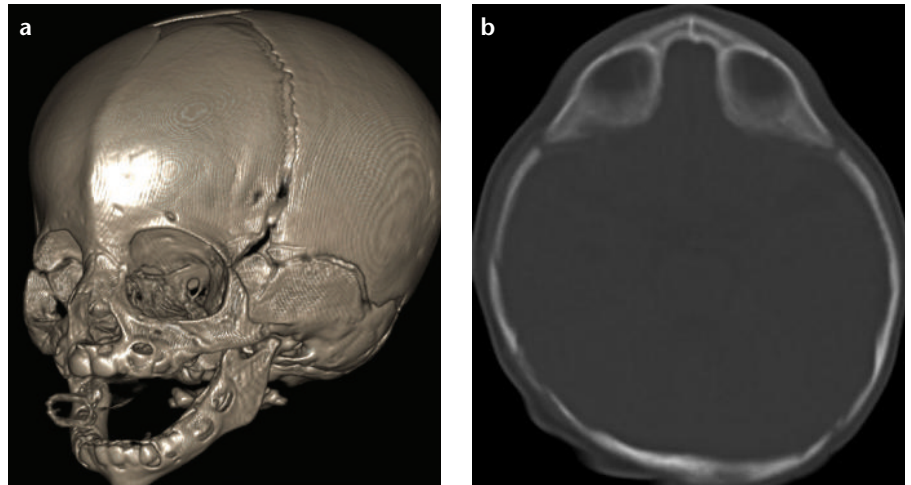
Scaphocephaly (or dolichocephaly) results from premature sagittal synostosis. There is increased growth following the direction of the sagittal suture (Virchow's law). This is the most common form of isolated synostosis (Fig. 9) (11).

Trigonocephaly is a bulging of the forehead due to fusion of the metopic suture before six months of age (1). Metopic suture fuses from the glabella to the anterior fontanelle. Anterior fontanelle ossification, hypotelorism, narrowing of the anterior cranial fossa and compensatory increase of the middle cranial fossa are observed (Fig. 10) (11).

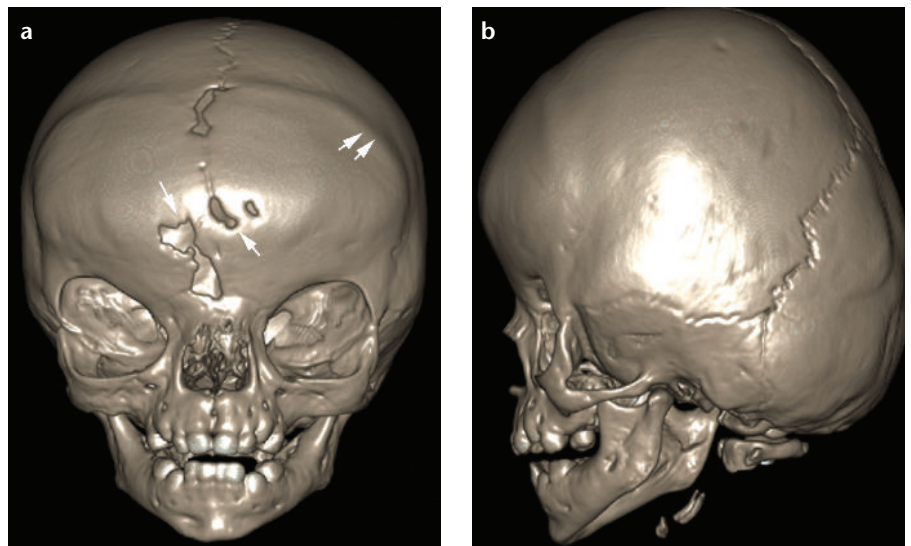
Oxycephaly or brachycephaly results from bilateral premature fusion of the coronal or lambdoid sutures. There is a flat and high forehead due to growth following the direction of the coronal suture. The transverse diameter of the skull is widened. Superior displacement of the lesser wing of the sphenoid bone causes the characteristic "harlequin eye" (1, 11). Brachycephaly is frequently seen with syndromic synostosis (e.g., Apert, Crouzon, Pfeiffer, craniofrontonasal syndromes) (Fig. 11).

### Congenital calvarial defects

Parietal foramina are paired parasagittal defects that result from delayed or incomplete ossification of the parietal bones (2). They are generally isolated but may be part of a syndrome. They are usually considered benign. Parietal foramina associated with an atretic cephalocele and symmetrical parietal meningoceles with



**Figure 10. a, b.** Trigonocephaly. Three-dimensional (a) and axial multiplanar (b) images of premature fusion of the metopic suture with hypotelorism, narrowing of the anterior cranial fossa, and compensatory increase of the middle cranial fossa.

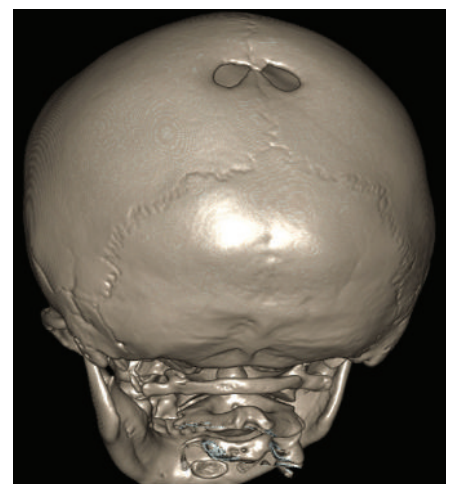


**Figure 11. a, b.** Craniofrontonasal syndrome and brachycephaly. Bilateral coronal synostosis with a bony ridge (a, double arrows). The calvarium is shortened in the sagittal plane and broadened in the transverse plane (b). Lateral displacement of the orbits and central defects between the frontal bones (a, arrows).

abnormal venous anatomy have been described (Fig. 12) (13).

Open sutures and anterior fontanelle can be due to elevated intracranial pressure (Fig. 13) or hypothyroidism and skeletal dysplasia—e.g., cleidocranial dysplasias, pycnodysostosis, and osteogenesis imperfecta.

Large anterior fontanelle can be associated with achondroplasia, congenital hypothyroidism, Down syndrome, rickets and increased intracranial pressure. The anterior fontanelle size is the average of the anteroposterior and transverse diameters. The average size of the anterior fontanelle



**Figure 12.** The parietal foramina is observed as paired parasagittal defects in parietal bones.



is 2.1 cm, and the median time of closure is 13.8 months (Fig. 14) (14).

### Calvarial bone fractures

Cranial fractures that are parallel or nearly parallel to the section orientation may be missed at interpretation of CT. A pediatric calvarium with multiple sutures and fontanelle makes the diagnosis more difficult. Three-dimensional VR and 3D MIP images are useful in these patients, and fractures and their extension can be assessed easily (Fig. 15) (12).

Cephalohematoma is a subperiosteal hematoma of the calvaria (2). They do not cross the midline. They generally resolve spontaneously and may calcify peripherally (Fig. 16). If they are not absorbed, they can ossify over the surface. Ossified cephalohematoma is a rare entity that requires surgical management and that can mimic osteoma in 3D images.

### Calvarial bone tumors

Three-dimensional CT may facilitate evaluation of lytic and sclerotic bone tumors. Three-dimensional CT is useful for preoperative and postoperative assessments of these patients.

Osteomas are the most common primary benign tumors of the calvaria. They are solid, nodular sclerotic lesions, which usually arise from the outer table (Fig. 17) (15).

Langerhans cell histiocytosis, epidermoid and dermoid cysts, meningioma, hemangioma, fibrous dysplasia, and metastases are other common lesions of the calvaria.

### Postoperative cranium

Three-dimensional CT is valuable for postoperative evaluation of surgery for craniosynostosis (Fig. 18). Burr holes, craniectomy defects and bone grafts may be evaluated using 3D VR images.

### Conclusion

Three-dimensional images should be included when reporting calvarial

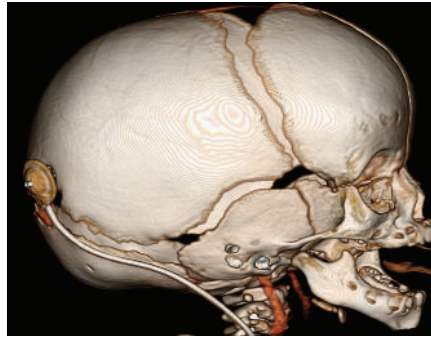


Figure 13. Large sutures and anterior fontanelle due to hydrocephalus.

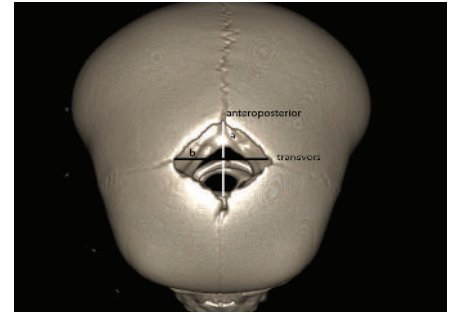


Figure 14. A large anterior fontanelle in a two-year-old male. Measurement of the fontanelle size  $(a+b/2)$  is seen.

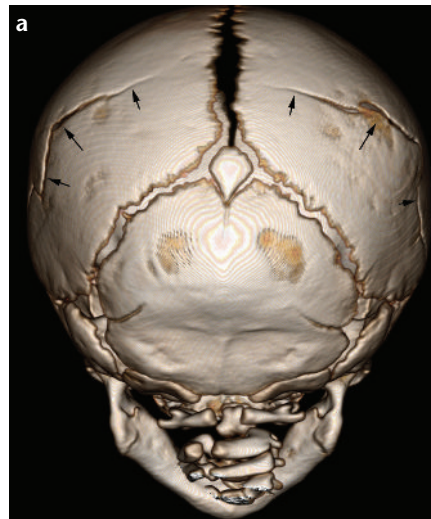


Figure 15. a, b. Bilateral parietal bone fractures (arrows) are seen in posterior view (a) and lateral view (b).

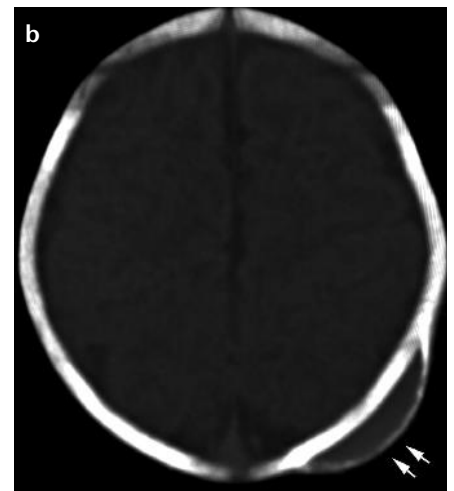
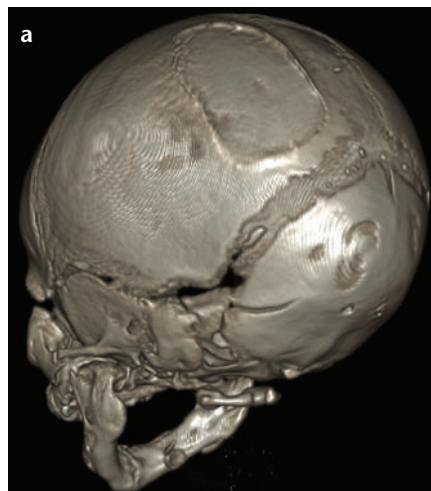
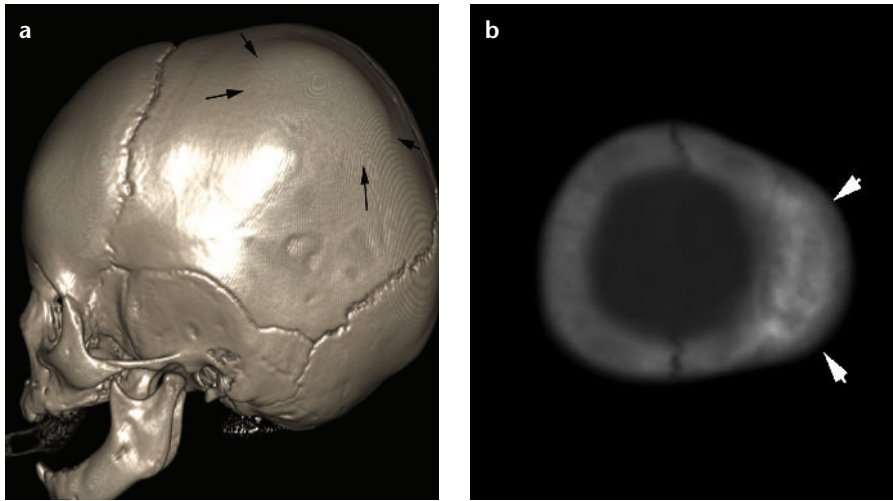


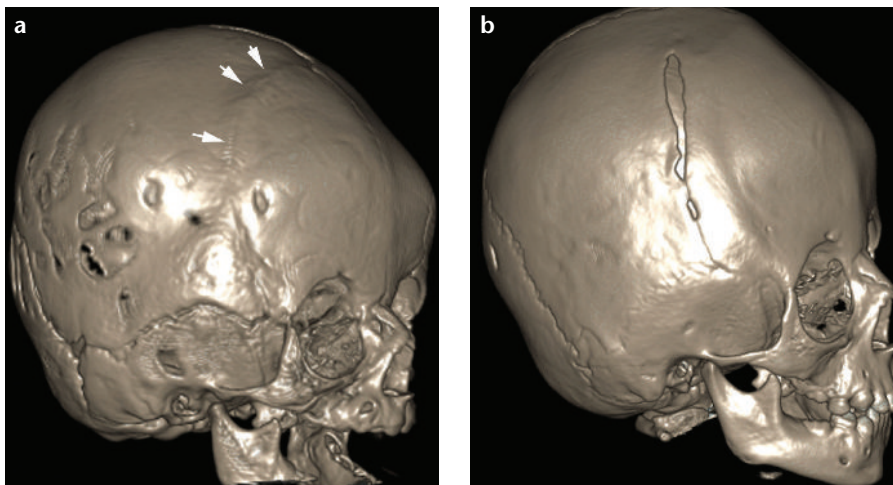
Figure 16. a, b. Cephalohematoma. Three-dimensional (a) and axial multiplanar (b) images show peripherally calcified subperiosteal hematoma (b, double arrows).

pathologies because 3D CT can aid in differentiating between a normal and an abnormal calvarium. It is particularly superior in the diagnosis of

craniosynostosis and fractures but also provides additional information regarding other pathologies.



**Figure 17. a, b.** Osteoma. Three-dimensional (a) and axial multiplanar (b) images show a left parietal osteoma (arrows).



**Figure 18. a, b.** Preoperative (a) and postoperative (b) images of a patient with anterior plagiocephaly due to fusion of the right coronal suture (a, arrows). Note that there are preoperatively increased convolutive markings in the parietal and occipital bones that result from increased cranial pressure.

#### Conflict of interest disclosure

The authors declared no conflicts of interest.

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