The calvaria in children, from normal variants to disease: A pictorial review

Poster No.: C-1884
Congress: ECR 2013
Type: Educational Exhibit
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Keywords: Education and training, Normal variants, Education, CT, Paediatric, Head and neck
DOI: 10.1594/ecr2013/C-1884

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Learning objectives

To describe the normal anatomy and embryological development of the calvarium.

To illustrate the wide variety of congenital and acquired lesions that affects the calvaria in children.

To review the low-dose Multidetector Computer Tomography (MDCT) technical and image reconstruction capabilities available for their accurate characterization.

To describe skull anatomical variants that may mimic disease and those radiological features that allow us to differentiate them.
Background

A wide spectrum of cranial vault congenital and acquired lesions is commonly encountered in children. They frequently show as calvarial deformities, bumps or defects, posing a significant source of concern for parents. Often clinical examination is not enough to reach a diagnosis, therefore imaging studies are crucial for the management of these patients.

Moreover, skull anatomical variants mimicking disease are common imaging findings and radiologists should be aware of them to avoid further diagnostic or therapeutic procedures.

IMAGING TOOLS

Plain film is the basic imaging tool, although it may be particularly inaccurate in the child under three months of age due to the low mineralization of the cranium. MDCT with its volumetric and post-processing capabilities (including Maximum Intensity Projection (MIP) and 3D Volume Rendering) is an excellent imaging modality to assess the skull. Furthermore, the bone high intrinsic contrast allows the development of low-dose protocols with tube voltage 80 and even 60 kVp. In addition, Computed Tomography allows evaluation of intracranial organs and detection of structural anomalies. Ultrasound (US) and Magnetic Resonance Images (MRI) are less useful tools regarding bone evaluation.

ANATOMY AND EMBRIOLOGY DEVELOPMENT Fig. 1 on page 29

The skull of the embryo starts developing between days 23 and 26 of gestation. It is divided into two distinct portions: the neurocranium, encasing the brain parenchyma, which develops from the surrounding mesenchyme, and the viscerocranium, which forms the facial skeleton and develops from the first three branchial arches. The neurocranium is further divided into the membranous neurocranium, which forms the cranial vault or calvarium, and the cartilaginous neurocranium, which forms the skull base.

The frontal, parietal and occipital bones and the squamous portion of the temporal bone form the calvarium. These flat bones are individually covered by periosteum, whose extracranial layer forms part of the scalp (pericranium) and is closely applied to the dura mater on the intracranial surface of the calvarium.
The frontal bone develops in membrane from dual ossification centres, one on either side of the midline.

The parietal bone ossifies in membrane from a single centre according to some authors and from two or more centres that fuse during the fourth month according to others. There is a slowing in the spread of ossification resulting in a notch in the postero-superior aspect of each parietal bone and in the widening of the sagittal suture at that level. This notch normally disappears in the fifth fetal month, but vestiges may be observed postnatally, in a number of bony defects (parietal cissures, small and large parietal fontanelles, obelial bones and small and large parietal foramina).

The occipital bone has a dual origin from membrane and cartilage. The membranous component gives rise to the interparietal segment (the superior portion of the occipital squama). The union of four primary centres around the foramen magnum forms the cartilaginous portion. These centres are the basioccipital anterior to the foramen magnum, the lateral or exoccipitals on either side of the foramen magnum, and the supraoccipital posterior to the foramen magnum (the inferior portion of the occipital squama).

In the newborn, the calvarial bones are unilaminar without diploe until the age of about four years. From this period on, two distinct cortical layers, the inner and outer tables, with an intervening marrow containing diploic space become more distinct and continue to do so into adulthood.

Calvarial bones are separated by bands of fibrous connective tissue, the sutures and synchondroses, which prevent premature bone separation and allow for uniform growth of the cranium during brain development. Sutures and synchondroses are prominent in the newborn, diminishing in width during the first two to three months. Their obliteration does not begin until the second to third decades.

Saggital suture separates both parietal bones. Metopic suture separates both ossification centres of the frontal bone; it usually disappears by the end of the second postnatal year, although it may persist throughout life in about 10% of cases. Coronal sutures separate frontal bones from parietal bones. Lamboidal sutures separate occipital and parietal bones. Mendosal sutures separate the lateral aspects of the membranous interparietal segment and the cartilaginous supraoccipital segment of the occipital bone. The exoccipital portions of the occipital bone are separated of the basioccipital portion by two prominents synchondrosis that usually disappears during the second or third year, and from the supraoccipital portion by the two innominate synchondrosis; they complete their obliteration at two to four years of age. At last the squamous sutures separate the parietal bones from the squamosa of the temporal bones.
Where the sutures intersect, they widen and assume the shape of fontanelles. The anterior fontanelle lies at the intersection of the sagittal, coronal, and metopic sutures and closes by the end of the second year. The posterior fontanelle lies at the intersection of the sagittal and lambdoid sutures and closes before the third month, occasionally, may be closed at birth. Two sphenoidal or anterolateral fontanelles lie at the intersection of the coronal, squamous and frontonasal sutures and two mastoid or posterolateral fontanelles lie at the intersection of the innominate synchondrosis and squamous and lambdoidal sutures. Closure of the fontanelles occurs clinically before it does radiographically.

The most significant growth of the skull occurs along the sagittal and coronal sutures.

At birth, the volume of the neurocranium is eight to nine times greater than that of the face. This ratio is 5:1 by two years, 3:1 at six years, and 2:1 in the adult.

The scalp covers the cranial vault and is formed by five layers: skin; superficial fascia or subcutaneous tissue, which contains blood vessels and nerves; muscle aponeurotic layer or galea aponeurotica, that consist in frontal and occipital bellies of the frontooccipital muscle; loose subaponeurótic layer, a space filled with loose areolar conective tissue crossed by emissary veins and, at last, the pericraneum. Fig. 2 on page 29

**SEMILOGICAL CLASSIFICATION OF CRANIAL VAULT NORMAL VARIANTS AND LESIONS IN CHILDREN** Fig. 3 on page 30

Calvarial lesions may be classified as malignant and benign, based on their biological activity or as congenital, tumoral, inflammatory, or traumatic, according to their histopathological features. In this pictorial essay we favor a semiological classification. Cranial vault lesions were discussed according to their clinical and radiological features as masses, defects, deformities, lines or abnormal skull density.

**SKULL DEFORMITIES OR ABNORMAL SKULL SHAPE**

**Normal variants**

- Bathrocephaly

Skull configuration caused by an outward bulge of the occipital squamosa, in the region of the mendosal suture. Probably a developmental variation of the mendosal suture, it is believed by some to represent postural deformity resulting from breech position. Typically resolves with skull remodelling and is of no clinical concern.
Craniosynostosis

Craniosynostosis is the premature closure of a cranial suture that can be the result of a primary developmental anomaly or secondary to external causes (teratogens, lack of brain growth, intrauterine compression, hematological, metabolic, or dysplastic bone disorders).

It is a common malformation with an incidence of 1 / 2100-2500 live births. 90% of cases are sporadic and isolated (a single suture affected), without any identifiable genetic mutation, but around 10% are associated with syndromes and other developmental abnormalities. When multiple sutures are involved we should strongly consider syndromic or genetic causes.

Craniosynostosis are usually identified early. The brain grows rapidly during the first few years of life, and patient skull progressively changes shape deviating from normal. The deformity of the vault depends on the suture involved: the growth of the skull follows the direction of this suture, and is arrested at right angles to it. **Fig. 4 on page 31**

Early diagnosis is essential for management, prevention of complications, and consideration for early surgical correction.

Signs of synostosis can be appreciated on plain radiograph, such as size and skull shape, sclerotic suture margins, absence of a suture, and beaten copper pattern, but further imaging is required for detailed surgical planning. Plain film interpretation of suture patency is particularly inaccurate in the very young (less than three months) as the mineralization of the skull is very low.

CT scans confirms the diagnosis, establish its extent, and exclude associated abnormalities (Chiari malformation, cervical spine abnormalities, hydrocephalus), an important item in the preoperative planning. Most cranial sutures are best assessed using 3D reconstruction of the images (diagnostic accuracy approaching 90%-100%). The obliterated suture is manifested by bone bridging across the suture or loss of architecture. However 3D reconstructions can overestimate the degree of fusion resulting in false positives (wormian bones can simulate bone bridges on 3D reconstruction), therefore it must always be supplemented with MIP reconstructions, which provide greater reliability of bone texture.

**MRI is superior to CT in parenchymal structural and abnormalities of the cranial junction assessment.**
The use of US is not established. A normal suture on US transverse plane is seen as a hypoechoic gap between two hyperechoic bone plates. From the suture opening, the dura can be seen as a faint echogenic line. The edges of a normal suture can meet edge to edge, be beveled, leveled, or overlapped. Obliteration of the hypoechoic gap, loss of the beveled edge, irregularity and thickening of the inner margin of the suture, and irregular anterior fontanelle, are all signs indicating potential suture fusion. Fig. 5 on page 32

*Isolated synostosis*

They are normally sporadic and associated with normal intellectual development. Cross-sectional imaging is needed to exclude underlying abnormalities, plan surgical procedures, and avoid misdiagnosis of nonsynostotic skull deformities.

- **Sagittal synostosis** Fig. 6 on page 33

It is the commonest form of isolated craniosynostosis and occurs more often in boys than girls. Patients may present with a dolichocephalic head shape, sagittal ridging and frontal and occipital bossing due to restricted growth in the biparieto-temporal areas and compensatory growth along the coronal, metopic and lambdoid sutures. CT shows focal or generalized absence of the sagittal suture.

- **Plagiocephaly**

Plagiocephaly refers to any flattening of the calvarium without denoting its aetiology and is preceded by terms describing its location and side.

It may be due to unicoronal or unilambdoid synostosis or to a benign positional deformation with normal sutures. In anterior plagiocephaly synostosis is most often the underlying cause, whereas in posterior plagiocephaly the cause is most commonly due to a nonsynostotic deformation.

Unicoronal synostosis is the second most common form of isolated craniosynostosis and occurs slightly more often in girls than boys. It results in plagiocephaly with a characteristic concave flattening of the forehead, elevated supraorbital ipsilateral margin and contralateral frontal bossing. This distortion results in a characteristic finding on coronal radiographs called the harlequin sign. The deformity affects the orbital position, resulting in outward deviation of the ipsilateral orbit and amblyopia. Fig. 7 on page 34

Unilambdoid synostosis is the least common form of nonsyndromic synostoses (4% of cases). It affects the posterior fossa and foramen magnum. Ipsilateral to the
synostosis, there is flattening of the occiput and compensatory bone growth of the squamosal suture and mastoid process. On the contralateral side, there is parieto-occipital bulging and compensatory overgrowth of the contralateral lambdoid and sagittal sutures. The calvarium is trapezoid in shape compared with parallelogram in deformational plagiocephaly. The midline of the foramen magnum is deviated toward the lambdoid synostosis. There is a windswept appearance of the skull with the posterior aspect of the vertex falling of to the contralateral side. The ear is pulled toward the synostotic side. Fig. 8 on page 35

- **Metopic synostosis Fig. 9 on page 36**

It represents approximately 10% of overall cases of craniosynostosis. When the metopic suture fuses before six months of age a trigonocephalic deformity (triangular shaped anterior cranial fossa) occurs. It is characterized by narrowing of the anterior cranial fossa with a pointed forehead or prominent midfrontal ridge. If the suture fuses later, then the deformity is minimal or not seen at all.

**Multisuture synostoses**

Multisuture craniosynostoses constitute a very small proportion of the nonsyndromic cases. They can be further subdivided into two-suture disease and multisuture synostoses. Multisuture synostoses have higher rates of complications and require complex surgery.

Bilateral coronal Fig. 10 on page 37 or lambdoid synostosis results in a brachycephalic skull, short in the antero-posterior diameter. The most severe form, often due to combined bicoronal and bilambdoid synostoses, is known as cloverleaf skull or Kleeblattschädel syndrome and is characterized by towering and narrow skull shape, with bulging temporal areas, shallow orbits, recessed supraorbital rims and hypoplasia of the basal frontal bones.

**Syndromic craniosynostoses**

Syndromic craniosynostoses differ from isolated synostoses in that they are often associated with other skeletal abnormalities (midfacial hypoplasia, cleft palate, hand abnormalities) and in an increased incidence of raised intracranial pressure (ICP) and hydrocephalus, optic atrophy, respiratory problems, disorders of speech and hearing in this patients. More than one hundred eighty syndromes have been described to date, including Crouzon, Apert, Pfeiffer and Muenkes syndromes. They present with multisuture synostoses that are nonspecific to each syndrome. The differential diagnosis relies on examining associated anomalies produced
in the face, hands and feet. Skull base, facial, and brain anomalies define the management of these patients.

• **Apert syndrome**

It is associated to one of the two mutations in FGFR2, involving Ser252Trp and Pro253Arg, two adjacent amino acids. Bicoronal craniosynostosis, midface hypoplasia, exorbitism, and hypertelorism occur. The symmetric severe complex syndactyly of fingers and toes is a distinguishing feature of Apert syndrome, which clinically differentiates it from other craniosynostotic syndromes.

• **Crouzon syndrome  *Fig. 11 on page 38***

Multiple mutations in the FGFR2 gene have been attributed to the syndrome of Crouzon. The triad of calvarial deformities, facial anomalies, and exorbitism characterizes it. Crouzon syndrome presents with bicoronal synostosis, exorbitism, hypertelorism, and midface hypoplasia. The lack of hand and foot anomalies differentiates it clinically from other synostotic syndromes. Other associated anomalies exist, but they are less severe than Apert syndrome.

• **Pfeiffer syndrome  *Fig. 12 on page 39***

Consists of craniosynostosis affecting multiple sutures, midfacial hypoplasia with severe exorbitism, and hypertelorism. Peripheral skeletal anomalies are common with broad and medially deviated great toes, broad thumbs, and soft-tissue syndactyly. A rare single recurring mutation in FGFR1 (Pro252Arg) and several different commoner mutations in FGFR2 have been identified.

• **Muenke syndrome**

The Pro250Arg in FGFR3 is the commonest mutation causing coronal synostosis. The hands and feet are affected in some cases but most abnormalities are not clinically significant. The importance of identifying limb anomalies (thimble-like middle phalanges, carpal or tarsal coalition or coned epiphyses) in combination with coronal synostosis, is that they strongly suggest the diagnosis of Muenke syndrome and warrant genetic testing for the Pro250Arg mutation. All patients should be tested for sensorineural hearing loss.

**Deformities mimicking craniosynostosis**

• **Postnatal deformational plagiocephaly  *Fig. 13 on page 40***

Postnatal deformational plagiocephaly results from external pressure after birth when an infant is consistently placed in the same position. The deformity may be unilateral or bilateral and symmetric or asymmetric, with one side substantially more flattened than the other.
Since the American Academy of Paediatrics recommended in 1992 that neonates and infants should be placed supine to sleep in order to decrease the incidence of sudden infant death syndrome, occipital plagiocephaly has increased alarmingly.

Anterior displacement of the ipsilateral ear and ipsilateral frontal and contralateral occipital bossing allow differential diagnosis with very uncommon unilateral lambdoid synostosis.

Severe cases may come to radiological evaluation to exclude lambdoid fusion.

- **Pseudoscaphocephaly**

It occurs when premature infants lay on the side of their heads in the neonatal intensive care for mechanical ventilation. Their heads become flattened and scaphocephalic.

### Traumatic deformities

- **Skull molding** [Fig. 14 on page 41](#)

During vaginal delivery, the fetal skull is often deformed as it passes through the birth canal. This skull deformity consists of posterior and superior displacement of the parietal bones and elevation of the occipital bone. Skull molding typically resolves within days.

- **Congenital depressions**

Congenital depressions are deformities of the calvaria due to mechanical factors that operated either before or during birth. They are usually satisfactorily visualized by direct inspection, but radiographs are often made in the search for associated fractures.

During labor, depressions of the calvaria are caused by excessive localized pressure on the head by the bony prominences in the maternal pelvis (sacral promontory, pubic symphysis, sciatic spines) or, less common, by the application of forceps to the fetal head (ping-pong ball depressions); these can be elevated by simple tangential digital pressure on opposite sides of the depression or by suction with a hand breast pump.

The term "faulty fetal packing" refers to concave depressions in the neonatal skull that are caused by prolonged extrinsic pressure from a malpositioned limb in uterus. Usually they are not associated with edema or hemorrhage of the underlying soft tissues. The skull deformity is not permanent and will resolve with time. [Fig. 15 on page 42](#)

Deformities has also been shown to occur from extrinsic pressure caused by uterine leiomyomas or amniotic bands.

- **Depressed fractures** [Fig. 16 on page 43](#)
A skull fracture is considered depressed when any portion of the outer table of the fracture line lies below the normal anatomic position of the inner table. Depressed fractures occur when objects with a large amount of kinetic energy make contact with skull over a fairly small area.

SKULL DEFECTS

Normal variants

- Accessory fontanelles

The most common (5-6% of newborns) is the small parietal fontanelle, (also named as the interparietal, sagittal, accessory, obelian or third fontanelle or fontanelle of Gerdy). Possibly it is more frequently in Down syndrome or associated with hypothyroidism. It consists on a round or diamond-shaped widening of the sagittal suture approximately 0.5 cm in size and located at the obelian region (about 2 cm in front of the posterior fontanelle in newborns). It is not known how often it leads to small parietal foramina. Occasionally it indents only one parietal bone, the so-called unilateral sagittal fontanelle.

Rarely, a metopic fontanelle can be present.

- Small parietal foramina Fig. 17 on page 44

Also called emissaria parietaria or foramina Santorini. They are two minute round defects, about 1 mm in size, located at the level of the obelion, one on each parietal bone, usually within 1-2 cm of the sagittal suture. Each foramen transmits an emissary Vein (Vein of Santorini), which connects epicranial occipital veins with the superior sagittal sinus and sometimes also a very small artery anastomosing a branch of the occipital with a branch of the middle meningeal artery. They probably represent a persistence of the most lateral aspects of the primitive parietal notch and occur in 60%-70% of adults, being unilateral in almost half of the cases.

It is not proven that once present they ever close spontaneously.

Abnormally large fontanelles or delayed fontanelles closure

They may arise in conjunction with suture spreading from elevated ICP, as part of skeletal dysplasias (acndroplasia, osteogenesis imperfecta, cleidocranial dysplasia), metabolic or endocrine disorders (hypothyroidism, rickets) or Down syndrome.
The delayed closure of the anterior fontanelle is thought to be secondary to delayed midline ossification of the metopic and sagittal sutures, allowing the anterior fontanelle to remain open into the adult life. Fig. 18 on page 45

**Large parietal fontanelle and large parietal foramina**

A large parietal fontanelle is a midline bony defect in the posterior interparietal region, which is larger than the usual parietal fontanelle and can reach great size. Fig. 19 on page 46 The larger defects are confluent with the posterior fontanelle and may be accompanied by bulging of the local soft tissues during crying. Probably represents a true anomaly in the ossification of the parietal bone. The differentiation between small and large parietal fontanelle is not clear-cut. The fate of the defect varies, being in most cases the precursor of parietal foramina of varying size, possibly depending on the size of the original bony defect. In rare cases the defect may persist as a midline parietal or sagittal foramen.

Large parietal foramina are two symmetrical oval or rounded parasagittal defects located one on each parietal bone, slightly anteriorly to the lambda. They can be up to 5 cm in size. They are closed by a fibrous membrane, which belongs both to the dura and the pericranium. The overlying scalp is normal, but rare cases have been reported in which it was the site of a congenital hairless defect.

They are said to represent an anomaly of ossification of the parietal bone of unknown cause, unrelated to small foramina. A familial incidence has been observed in a number of cases with a suggested dominant mode of inheritance. The defect may occur as an isolated lesion or in association with other malformations. Fig. 20 on page 47

**Cranium bifidum or cranioschisis**

It is an abnormal osseous defect in the skull due to defective induction of bone, pressure erosion by a mass or cyst, or failure of one of the sites of primary neural tube closure.

If intracranial tissue herniated through it results in an encephalocele. They are classified as meningoencephalocele (meninges, cerebrospinal fluid, and brain), meningocoele (meninges and cerebrospinal fluid), atretic parietal cephalocele (meninges and neural rests), and gliocele (cerebrospinal fluid filled glial-lined cyst), based on the neural elements they contain. Encephaloceles occur in 1 of every 4,000 live births and are most commonly occipital in location (75% of cases) being frontoethmoidal in 15% of cases and basal in 10%. There are often significant associated intracranial anomalies. Occipital encephaloceles may be associated with Chiari or Dandy-Walker malformations and callosal or migrational anomalies.
Surgery is the treatment used for encephaloceles, and MRI is the best imaging modality for defining the contents of an encephalocele prior to surgery. MRI with MR venography allows demonstrated venous involvement commonly associated to occipital encephaloceles.

High-resolution CT is superior in defining the usually midline bone defect.

**Cleidocranial dysplasia**

It is an autosomal dominant syndrome affecting membranous bone, characterized by poor or retarded mineralization of the skull with widening of the fontanelles, broad lateral cranial diameter and multiple Wormian bones along the lambdoid sutures; closure of the sutures and fontanelles occurs late. Another associated skeletal anomalies include absent or hypoplastic clavicles, a widened pubic symphysis, multiple spinal anomalies, and hypoplastic middle and distal phalanges. Hearing loss occurs in 38% of cases.

**Delayed sutural closure**

It can be due to any cause of increased ICP, achondroplasia, congenital hypothyroidism, Down's syndrome or rickets. Fig. 21 on page 48

**SKULL LINES**

**Normal variants**

- Wormian bones
- Intrasutural bones Fig. 22 on page 49

They are small and irregular intrasutural bones, most frequently found around and in the lambdoid suture. Its incidence varies ranging from around 10% (Caucasians) or 40% (Indians) to 80% (Chinese). Wormian bones are considered abnormal or clinically significant when there are more than 10 in number, measuring greater than 6 mm by 4 mm, and arranged in a mosaic pattern. In this case they can be seen in a variety of conditions particularly bone dysplasias, such as osteogenesis imperfecta, pyknodysostosis, and cleidocranial dysostosis or in disorders, such as Down syndrome, rickets, hypothyroidism, and hypophosphatasia.

- Intrafontanellar bones
Much less frequent than intrasutural bones

- **Intraparietal or inca bone** *Fig. 23* on page 50

It is a solitary wormian bone within the posterior fontanelle that results from division of the supraoccipital portion of the occipital bone into two parts by the mendosal suture, the superior part arising from membranous bone and the inferior part from cartilage.

- **Epipteric bone or pterion osicle**

It is a wormian bone within the anterolateral fontanelle (pterion).

- **Anterior fontanellar bone or bregmatic bone** *Fig. 24* on page 51

It is a rare wormian bone within the anterior fontanelle. It is clinically important as it can mimic premature closure of the cranial sutures by giving the appearance of a "closed fontanelle" and may be confused with a cranial fracture. It is thought to not interfere with cranial growth.

- **Obeliac bone**

Occasionally there is a fontanellar bone within a small or large parietal fontanella. In some instances, two or more Wormian bones appear within the defect.

- **Accessory sutures**

The parietal and occipital bones are common regions for accessory sutures because of their multiple ossification centres.

According to some authors the parietal bone ossifies from two centres oriented craniocaudally. If these centres fail to fuse, anomalous horizontal parietal sutures can results. Other parietal sutures may be vestiges of the fetal parietal notch, presenting as thin clefts in one or both parietal bones extending outwards from the sagittal suture for 1 cm or more.

The occipital bone has a more complex development. The supraoccipital ossification centre contains a midline occipital fissure that can, sometimes, persist postnatally.

Accessory sutures are usually bilateral and symmetric, especially in the parietal bones. Occipital accessory sutures can be complex and multiple but are also frequently bilateral. They show a zigzag pattern with interdigitations and sclerotic borders similar to main calvarial sutures.

Its importance lies in that they may be misdiagnosed as fractures (see lineal fractures)
• **Diploic and vascular markings**

The diploic veins of Bresched lie in large and irregular channels shown in plain radiographs of the cranial vault as strips of diminished density with very variable size and visibility that run in all directions. Also do the grooves of the internal aspect of the calvaria for arteries and veins. The largest and heaviest vascular markings are due to the bonny thinnings over the venous sinuses of the dura mater. They must be taken in account to avoid confusion with linear fractures.

**Traumatic lesions**

• **Linear fractures Fig. 25 on page 52**

Linear fractures and accessory sutures can be differentiated in most cases by observing its radiological characteristics; knowledge of the normal anatomy, development, and timing of sutural closure are also necessary to decipher the varied and complex nature of these accessory sutures especially in the occipital region.

Usually, simple non-depressed skull fractures are unilateral lineal pattern sharp lucencies with non-sclerotic edges that may show some degree of displacement. If they extend into a major suture, there could be widening of the fracture line as it approaches the suture or associated diastasis of the adjacent synchodrosis or suture. High impact fractures can cross suture lines, extend from one major suture to another or be bilateral, but in this cases will often show comminution, depression, and marked asymmetry. At last, soft tissue swelling or hematoma is frequently associated with acute skull fractures.

In contrast, accessory sutures, usually bilateral and symmetric, show a zigzag pattern with interdigitations and sclerotic borders similar to major calvarial sutures. They join and merge with major sutures.

Knowledge of the normal anatomy, development and timing of sutural closure are also important in the evaluation of questionable fractures. The occipital and innominate sutures are no longer apparent by age four while the mendosal suture completely fuses by six years of age. An example of an accessory suture that can be misleading is the normal persistent occipital suture. It extends from the dorsal aspect of the foramen magnum and can appear wide and sharp. However, it should extend no more than 2 cm from the edge of the foramen magnum. A longer fissure would be inconsistent with its normal embryogenesis and therefore represents a fracture. In some cases where the lucency is shorter than 2 cm, the age of the patient would help in deciding if this is a fracture or just a sutural remnant: a persistent lucency beyond age four is indicative of a fracture. **Table 1 on page 53**
However, in difficult cases, it is prudent to request for a follow-up study to look for signs of healing.

- **Leptomeningeal cyst: growing fractures**

They are a late and rare complication of head traumas (0.6% of skull fractures) that usually occur before the age of three years (90% of cases). Commonly affect the parietal bone.

There are two different forms: growing fractures and intradiploic arachnoid cysts. In both, there is a dural tear with an intact arachnoid that allows the herniation of cerebrospinal fluid (CSF) and, occasionally, brain parenchyma through it, interposing between the fractured fragments. This prevents osteoblastic migration and healing of the fracture. Fig. 26 on page 54

In growing fracture a progressive widening of the fracture line (> 4 mm) occurs, due to erosion of CSF pulsatile pressure. The fracture involves both the inner and outer table. It presents as an enlarging and sunken palpable cranial defect.

An intradiploic arachnoid cyst erodes the inner table rather than the outer table and it presents as an enlarging pulsatile mass.

Plain radiograph shows a persistent diastatic fracture.

On cross-sectional imaging, intracranial tissue or a leptomeningeal cyst may be seen to. There are frequently associated encephalomalacic changes of the underlying herniating brain tissue. CT detects both, the skull defect and the leptomeningeal cyst with CSF-like density extending between the edges of bone.

On MRI the cyst is isointense with CSF on all sequences and communicates with the subarachnoid space. Encephalomalacia of the underlying brain tissue appears as low density on CT and as T1 and T2 prolongation on MRI.

Early diagnosis is crucial, since brain damage is progressive in the absence of surgical treatment

- **Sutural diastasis**

Pathologic diastasis of the cranial sutures may be secondary to trauma, tumour deposits within the sutures, increasing ICP (hydrocephalus) or metastatic neuroblastoma. Rapid increases in ICP particularly in an infant may produce sutural diastasis. The coronal
suture is usually the first to demonstrate diastasis, and there may be associated bulging of the anterior fontanelle.

VARIATIONS IN SKULL DENSITY

Generalized decreased density

Bony dysplasias

- Osteogenesis imperfecta

It is a bone dysplasia due to mutations within the COL1A1 and COL1A2 genes, resulting in defective quality or quantity of type-I collagen. The clinical and imaging features depend on its type (I-VII) and severity. Diagnostic features of osteogenesis imperfecta include osteopenia, cortical thinning, reduced ossification of the skull, multiple Wormian bones along the lambdoid suture, and increased risk of fractures. Macrocephaly may be present secondary to a communicating hydrocephalus.

- Achondrogenesis

Achondrogenesis is a group of severe disorders that affect cartilage and bone development. It can be manifested as severe thinning of the calvaria and decreased calvarial density. A diagnosis of achondrogenesis is more likely when the patient also exhibits areas of absent ossification in the axial skeleton, micromelia and hydrops.

Metabolic disorders

- Hypophosphatasia

It is an inherited heterogeneous disorder caused by a deficiency of alkaline phosphatase, typically causes reduced ossification of the skull and vertebrae and may result in isolated plates or islands of unusually thin calvarial bone. The skull may be boneless and represented by a caput membranaceum. Other skeletal changes include short tubular bones poorly and irregularly ossified and frayed metaphyses that resemble rickets.

- Menkes syndrome

It is a rare X-linked recessive disorder of copper metabolism. Key differential features for Menkes syndrome are osteopenia, mental retardation, micrognathia, metaphyseal spurs most obvious in the femora, urinary tract abnormalities and high serum copper levels.

Localized decreased density
Normal variants

- **Digital or convolutional markings**

Decreased density areas of the calvaria, separated by normal density strips, which correspond with the location and configuration of cerebral convolutions and probably formed by localized pressure of the pulsating brain on the inner table of the neurocranium. They occur after sutural closure (three to seven years old) and can be very conspicuous in case of increased ICP.

Lacunar skull

Lacunar skull develops during fetal life and is present at birth. It is practically always associated with neural tube defects, especially myelomeningocele with Chiari II malformation, and less commonly with encephalocele. All patients under three months old with a meningomyelocele or encephalocele have some radiological evidence of lacunar skull.

The cause is not known but is probably a displasia of the calvarial membranous bone, more prominent in the parietal and upper occipital bones, characterized by localized well-defined oval lucent areas (lacunae) due to thinning of the inner table and diploic space, that correspond to nonossified fibrous bone. The lacunae are bounded by normally ossified bone. It is not related to increased ICP, as it is found in heads that are normal or small in size without evidence of hydrocephalus.

It must be distinguished from the true convolutional markings that occur later (after suture closure) and tend to appear first in the posterior and lower lateral portions of the calvaria.

Lacunar skull usually resolves spontaneously by age six months.

**Osteomyelitis**  

Osteomyelitis of the skull is rare in children. It can be due to bacteremia, trauma or direct extension of infection from sinus or scalp cellulitis. During the early stages of bone infection, the radiographic findings are negative. Later, when areas of inflammatory necrosis of sufficient size develop, they can be identified radiographically as areas of diminished density of variable size, shape, and position involving the inner and outer tables. In chronic osteomyelitis, sclerotic changes of the affected bone are usually present.
Generalized increased density

Bony dysplasias

- **Osteopetrosis**

It is an inherited heterogeneous group of osteosclerotic bone dysplasias in which the entire skeleton is unusually dense. Impaired bone resorption results in abundant osteoid and narrow and fibrotic medullary spaces. Sclerosis initially affects the basal bones and later the calvaria which become dense and thick. Bones are fragile and prone to fracture and show a high susceptibility to osteomyelitis. Neural and vascular foramina are narrow, causing cranial nerve palsies by neural compression.

The facial bones are usually less dense.

- **Pyknodyostosis**

It is a bony dysplasia characterized by thick calvaria, wide lambdoid sutures and fontanelles, multiple wormian bones, short limbs, hypoplasia of the mandible and an obtuse mandibular angle.

- **Craniodiaphyseal dysplasia**

It is a congenital bone dysplasia characterized by severe osteosclerosis with overgrowth of the skull, facial bones, and mandible. Obliteration of the paranasal sinuses and basal skull foramina and thickening of the diaphyses appear later. Sclerosis in the remainder of the skeleton is less marked.

Hemolytic anemias

In congenital haemoglobinopathies, such as thalassemia or sickle cell disease, and hereditary spherocytosis the bone marrow becomes hyperplastic. This causes a whitening of the diploic space owing to external displacement of the outer table.

Localized increased density

Bony dysplasias

- **Frontometaphyseal dysplasia**

It is an X-linked dominant bone dysplasia characterized by prominence of the supraorbital ridges, restricted thoracic expansion, sternal deformity, and joint contractures. The manifestations are more severe in males and variable in females. Radiologic findings
include sclerosis limited to the frontal bone and the skull base, premature synostosis of the sutures, anterior mandibular spur, widening of the metaphyses, arachnodactyly, "coat hanger" configuration of the ribs, and coxa valga.

- Craniometaphyseal dysplasia

It is a dominant inherited bone dysplasia in which the sclerosis is confined to the frontal bone, nasion, and mandible; metaphyseal expansion is a late feature.

**Rickets**

Some rachitic patients present with regional thickening of the calvaria caused by excessive amounts of poorly mineralized bone heaped up on the outer tables of the parietal and frontal eminences. Clinically they may produce local bosses. During the active phase of the disease, when the mineral content of bone is reduced, the thickenings are not well visualized radiographically, but with healing the calcium contents increase and the hyperostosis become more evident.

**CALVARIAL MASSES (WITH OR WITHOUT ASSOCIATED LYtic BONE LESIONS)**

**Normal variants**

- Prominent external occipital protuberance [Fig. 28 on page 56](#)

It is a normal variant clinically palpable as a hard lump in the occipital midline.

**Vascular lesions**

- Haemangiomas

Primary hemangiomas of the skull are benign lesions that develop in the diploic space and consist of dilated blood vessels with fibrous septa. They can be found in any location, but they are most often seen in the frontal and parietal regions.

Hemangiomas can manifest as palpable masses or be incidentally detected during an imaging evaluation.

Plain radiography is often the initial technique used, and it is useful for showing the exact location of a lesion.
CT is optimal for characterizing the bony features of the lesion and the lesion matrix, however, intradiploic and intracranial extensions and involvement of adjacent neurovascular structures are more readily detected with MRI.

Angiography assists in the pre-surgical evaluation and possible embolization of the tumor.

- **Venous malformations**

Venous malformations include a wide spectrum of dysmorphic and congenital venous lesions that are characterized clinically by a soft and nonpulsatile mass, often of bluish color.

Up to 40% of venous malformations occur in the head and neck.

MRI demonstrates a hyperintense mass on T2-weighted images, with occasional septation and variable enhancement. Phleboliths, which appear as a focal signal void, are a relatively specific characteristic. As a low-flow lesion, the flow voids demonstrated with proliferating hemangiomas or high-flow arteriovenous malformations are not seen in venous malformations. MRI is essential to allow evaluation of the underlying major venous sinuses prior to any planned surgical intervention.

Treatment typically involves some combination of sclerotherapy and surgical removal.

- **Sinus pericranii**

It is a soft, bulging, fluctuant mass, often of a red blue colour and less than 1.5 cm in diameter, located in the scalp, over the region of the sagittal or transverse sinuses in the frontal midline or parietal regions that may be associated with an underlying bony defect of the calvaria. Typically expands with an increase in ICP and compresses by direct pressure on the lesion.

Sinus pericranii results from an abnormal communication between intracranial venous sinuses and extracranial nonmuscular dilated scalp veins through diploic veins. It is often congenital, although it may be spontaneous or traumatic in origin.

Usually its significance is only cosmetic but common symptoms include headaches, sensations of pressure or fullness, or local pain. In rare cases, severe symptoms such as bradycardia, bradypnea, or hearing loss have been described.

The varicosities tend to increase in size slowly, although there has been a report of spontaneous regression.
Treatment, when necessary, usually consists of ligation of the communicating veins and surgical removal of the sinus itself.

On plain film examination there is irregular thinning of the bone without sclerosis.

On CT the soft-tissue mass enhances intensely, unless thrombosis of the communicating vein has occurred. There is scalloping of both or one table of the skull vault and a channel through the skull may be seen.

MRI is the best imaging modality for it characterization. There is a mixed intensity lesion with signal void areas within the lesion that enhances homogenously. Communication with a sinus may be demonstrated.

Interrogation with colour Doppler can demonstrate blood flow from the dural venous sinus to the extracranial lesion.

- Lymphangiomas

Lymphatic malformations of the head and neck are rare. They develop from lymphatic sacs that fail to communicate with the remainder of the lymphatic system. Macrocystic lymphatic malformations (cystic hygromas) have characteristic MRI features, manifesting as multiseptate cystic masses, often with intracystic haemorrhage or fluid levels. Treatment typically involves some combination of sclerotherapy and surgical resection.

Congenital inclusion cysts Fig. 29 on page 57

Congenital inclusion cysts are thought to represent the persistence of ectodermal elements at sites of suture and neural tube closure or diverticulation of the cerebral hemispheres. The majority of epidermoid and dermoid cysts are congenital, and are rarely due to traumatic or iatrogenic implantation. They may be located in the scalp, in the diploic space, or between the internal surface of the inner table and the dura, most frequently in midline (anterior fontanelle, glabella, nasion, vertex, and subocciput) and frontotemporal locations, followed by parietal locations. Sutures commonly affected include the frontozygomatic, sphenofrontal, sphenosquamosal, squamosal, coronal, lambdoid, and parietomastoid sutures. Epidermoids are typically lateral in location, and dermoids are usually found in the midline.

Dermoid cysts are most commonly seen in newborns and infants up to three years old and contain ectoderm and skin elements (hair, sebaceous and sweat glands).
lined by epithelium, whereas the less common epidermoid cysts contain ectoderm but no skin elements.

They are usually benign and grow slowly. If they protrude into the cranial cavity, they may be the source of cerebral symptoms. In the majority of cases, the lesions disappear within a few years of discovery; a transient period of moderate enlargement occurred prior to spontaneous resolution.

CT shows a well defined round or oval shaped, non-enhancing oval mass, which, if grows within the bone or impinge on it, causes remodelling, expansion and erosion of the inner and outer tables, leading to a lytic bone lesion with smooth sclerotic margins. Its attenuation varies depending on content (fat attenuation with dermoid cysts, that may have foci of peripheral calcification within them, and fluid attenuation with epidermoid cysts). Unlike epidermoids, dermoids usually have a thick wall that enhances with intravenous contrast media.

The signal intensity at MRI depends on the contents of the cyst and may range from pure fluid signal intensity (hypointense on T1-weighted images, hyperintense on T2-weighted images) in an epidermoid cyst, unless very occasionally there is fat or hemorrhage within the cyst, to a more complex signal intensity (hyperintense on T1-weighted images, hypointense on T2-weighted images) in a dermoid cyst. Epidermoid cysts typically have bright signal intensity on isotropic diffusion-weighted MRI.

Cephaloceles (see cranium bifidum)

Traumatic lesions

Haematomas

• Caput succedaneum Fig. 30 on page 58

It is a subcutaneous fluid collection, which results in a local swelling of the scalp, usually located at the vertex and due to pressure on the presenting head, recognized at birth and disappearing after a few days. As opposed to cephalohematoma it can crosses suture lines. Contents of the caput (oedema fluid and blood) cast a shadow of water density that disappears without residual bone production or destruction. In rare cases, the hemorrhagic oedema can be so massive that it can be associated with shock. In such cases, intracranial hemorrhagic is often present.

• Cephalohematoma Fig. 30 on page 58
Cephalohematoma is a traumatic subperiosteal accumulation of blood of the calvaria, usually secondary to birth trauma and particularly related to forceps delivery. Its prevalence is close to 1%-2% in spontaneous vaginal deliveries and 3%-4% in forceps- or vacuum-assisted deliveries. It is almost twice as common in males than in females, and is more common in children of primiparous mothers. Haemorrhagic disease of the newborn used to be an added predisposing factor.

Fine linear fractures of the underlying bone, of no clinical significance, may be found on the sites of cephalohematomas and are thought to be the principal cause of the bleeding of the periosteum.

The pericranium and the sutures bound it, so it cannot cross sutures or midline (this restriction distinguishes it from subgaleal hematoma which does cross the midline deep to the galeal aponeurosis).

Cephalohematoma manifests as unilateral or bilateral firm, tense, soft-tissue mass, more frequently over the parietal or occipital bone, which characteristically extend over the entire surface of the affected bone and is sharply limited at the edges of the bone where the periostium is bound tightly to the membranous tissue of the sutures. It usually increases in size after birth, and resolve spontaneously by a few weeks to 3-4 months of age. Smaller bleeds may not be detected at or soon after birth and may present later as incidentally discovered head lumps. Although cephalohematomas are not usually of clinical significance, they may present a challenge for the clinician because they occasionally become infected, requiring drainage and antibiotic therapy.

In cases where the hematomas fail to resolve, progressive subpericranial osteogenesis results in a calcified cephalohematoma. Near the end of the second week of evolution, new bone begins to form under the elevated periostium; it appears first at the margins of the cephalohematoma, but soon the entire blood tumour is overlaid with a complete shell of bone. The incidence of calcification of cephalohematomas has been reported to occur in 3-5% of all cephalohematomas. Fig. 31 on page 59

They rarely require imaging, as the history and clinical examination suffice.

Plain film, US, CT or MRI may be used to show crescent-shaped lesions adjacent to the outer table.

At MRI, signal intensity typically follows that of subacute haemorrhage (hyperintensity on T1- and T2-weighted images) but may vary depending on the stage of the haemorrhage.
CT is the definitive standard for assessment of calcified cephalohematomas. In cases that present late (three-six months), the cephalohematoma feels hard, and both plain radiography and CT show a uniform, densely calcified mass that is located immediately adjacent to the outer table and is contained within the periostium. The imaging findings persist long after the clinical have disappeared. The outer table remains thickened, as a flat, irregular hyperostosis for several months and is gradually resorbed. In some cases, the space between a new shell of bone and the outer table remains widened for many years, and the space originally occupied by the haematoma becomes filled with normal diploic bone. In other cases, large and small cysts-like defects persist in the site of cephalohematoma for months. During the evolution of a cephalohematoma, a mixed picture of erosive changes and periosteal reaction can be worrisome, especially in the absence of a good clinical history. Occasionally, infantile cephalohematoma persist into adult life, when large segments of bone production and destruction may still be visible in the calvaria: the cephalohematoma deformans.

- **Subgaleal haemorrhage** Fig. 30 on page 58

Subgaleal haemorrhage may simulate cephalohematoma and also caput succedaneum. The swelling in the scalp, often over parietal region, usually are associated with underlying fractures and. It is due to the accumulation of blood beneath the aponeurotic layer, so that is not confined by the cranial sutures and the collection extends through them, in contrast to cephalohematomas.

Trauma from obstetric forceps is also a common cause. In older child it may result from hair-pulling stress in cases of child abuse or accidental entrapment of long hair in mechanical equipment. Occasionally it occurs without antecedent trauma in patients with hematologic abnormalities such as platelet defects.

**Fractures**

- **Leptomingeal cyst: Intradiploic arachnoid cyst** (see leptomeningeal cyst: growing fractures) Fig. 26 on page 54

**Infections**

- **Pott puffy tumour**

It is a subperiosteal abscess and osteomyelitis of the frontal bone usually due to direct spread from frontal sinus infection. Bacteria tumour usually reflects species responsible for community-acquired chronic sinusitis (streptococci, staphylococci, and anaerobic bacteria). The infection may spread as a thrombophlebitis from the frontal sinus through the diploic veins, involving the intracranial space with consequent epidural or subdural empyema, meningitis, brain abscess, and venous sinus thrombosis.
Contrast-enhanced CT or MRI is needed to evaluate for possible intracranial complications.

Surgical drainage associated to resection of infected bone and the granulation tissue in the scalp and prolonged antibiotic therapy remains the mainstay of therapy.

Neoplasms

Benign neoplasms

- **Osteomas**

Osteomas are the most common benign calvarial tumor. They are typically slow growing tumors that are formed by mature bone tissue. They are seen as solid, sclerotic, well-demarcated, nodular density on CT. They use to be less than a centimetre and arise from the outer table.

- **Fibrous dysplasia**

Fibrous dysplasia is a common skeletal lesion in adolescents and young adults that may affect the calvarium in both, the monostotic (70-80 % of cases) and polyostotic forms.

It is characterized by replacement of normal bone marrow by fibro-osseous tissue causing expansion of the diploe. Recent studies regard it as a benign non-encapsulated neoplasm. Calvarial fibrous dysplasia commonly crosses bony sutures, and, as such, monostotic lesions may involve multiple calvarial bones. Most small calvarial lesions are asymptomatic and are identified incidentally. More expansile lesions usually grow outward and present with firm swelling. Rarely, polyostotic fibrous dysplasia may be associated with McCune-Albright syndrome and Mazabraud syndrome. Malignant transformation of fibrous dysplasia occurs very infrequently (0.4 -4 % of cases).

On CT scanning three subtypes of fibrous dysplasia are recognized and in order of disease activity are cystic, pagetoid, and sclerotic types. The typical CT appearance is an intradiploic, expansile lesion with a characteristic "ground glass" homogeneous pattern (pagetoid type), in which the outer table is more prominently affected than the inner table, thus avoiding intracranial pressure effects.

On MRI signal intensity varies with regard to the amount of fibrous tissue and osseous matrix in the lesion. The most frequent type is hypointense on T1- and T2-weighted sequences with high T2 signal in areas of increased pathologic activity. Enhancement
after contrast media is variable, with homogeneous, central, or peripheral patterns described.

Radionuclide bone scintigraphy is useful to exclude polyostotic disease.

- **Langerhans cell histiocytosis** [Fig. 32 on page 60]

Langerhans cell histiocytosis is a systemic disease with a wide spectrum of presentation: Hand-Schuller-Christian's syndrome; Letterer-Siwe disease and eosinophilic granuloma. The last one is the most frequent (80%) and least aggressive form, limited to bone or lung and often monostotic.

Eosinophilic granulomas typically present during the first decade of life and may be solitary or multiple. The skull is a common site of involvement, especially the temporal and parietal bones, with typical "punched-out" osseous lesions; they begin in the diploe and invade the inner and outer tables.

On plain radiographs, it presents as a lytic skull lesion with a beveled edge or sharp and serrated margins, and without reactive sclerosis or periosteal reaction. A hole-within-a-hole appearance may also be seen because of uneven erosion of the inner and outer tables of the skull.

On CT it appears as an enhancing soft-tissue mass with bone erosion; the margins may be well defined but with beveled edges because of asymmetric destruction of the inner and outer table. Peripheral sclerosis is seen only in the healing period. Occasionally a button sequestrum, may be present.

On MRI, it is hyperintense on T2 weighted images, has variable signal intensity on T1 weighted images and enhances in post-contrast images. Edema and inflammation around the lesion may be present.

**Malignant neoplasms**

- **Melanotic progonoma**

It is a rare tumor of the skull. It usually begins during the first year of life as a movable scalp nodule that subsequently invades the bone and becomes fixed to it, often adhering to the dura. The bone is destroyed but reactive spicules develop internally and externally, producing a sunburst appearance.

- **Metastatic neuroblastoma** [Fig. 33 on page 61]
Neuroblastoma is the most common extra-cranial solid tumour in children. 90% occurs under the age of ten years. Bony metastases have been reported in 34-74% of cases with long bones and lateral orbits walls, manifesting clinically as two "black eyes" ("raccoon sign"), being the most common sites. Skull base and calvarium are also frequently affected.

Four patterns of calvarial involvement have been described:

1. Dural involvement with secondary sutural diastasis. Dura mater is resistant to tumour penetration so that tumour grows as a plaque-like epidural deposit that takes off the dura away from the inner table and may cause intracranial mass effect and induce sutural widening. As tumour spread is limited by tight dural anchorage to adjacent suture margins, the consequent pressure is the mechanism that cause the sutural diastasis.

2. Sunburst ("hair on end") or linear periosteal reaction.

3. Lytic bone defects multiple and poorly defined which may coalesce.

4. Thickened bone which leads to focal calvarial hyperostosis.

On CT shows a characteristic appearance of a mass or masses growing inward from the bone with spicules of bone radiating centripetally within them. The metastases can grow into the intracranial space mimicking an intracranial mass. Coronal images help to identify a dural or extradural origin. The lesions enhance markedly on CT and on MRI examination.

On MRI metastases appear as soft-tissue masses of high T2 signal intensity that enhance on fat-suppressed T1-weighted images originating within the calvarium. The elevated periosteum will also enhance and may be a helpful sign in differentiating metastatic tumour from normal haematopoietic marrow of childhood.

Bone scintigraphy will confirm or exclude multiple sites of involvement.

- **Leukemia.**

Characterized by localized areas of bone destruction surrounded by preserved bone. Associated features include infiltration of the appendicular skeleton and hepatosplenomegaly.
Fig. 1: Anatomy of normal calvarium and CT lateral and superior views 3D reconstructions.

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**Fig. 2:** Scalp layers

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**SKULL DEFORMITIES OR ABNORMAL SKULL SHAPE**

- **Normal variants**
  - Batrachcephaly
- **Craniosynostosis**
  - Isolated craniosynostosis
  - Sagittal synostosis
  - Plagiocephaly
  - Metopic synostosis
  - Multisuture synostosis
  - Syndromic craniosynostosis
  - Apert syndrome
  - Crouzon syndrome
  - Pfeiffer syndrome
  - Muenke syndrome
- **Deformities mimicking craniosynostosis**
  - Postnatal deformational plagiocephaly
  - Pseudoplaigiocephaly
- **Traumatic deformities**
  - Skull molding
  - Congenital depressions
  - Depressed fractures

**SKULL DEFECTS**

- **Normal variant**
  - Accessory fontanelles
  - Small parietal foramina
- **Abnormally large fontanelles or delayed fontanelle closure**
  - Large parietal fontanelle and large parietal foramina
  - Cranium bifidum or craniocriosis
  - Cleidocranial dysplasia

**SKULL LINES**

- **Normal variants**
  - Wormian bones
  - Intracranial bones
  - Intracranial fontanelles
    - Intraparietal or inca bone
    - Epiphragmatic bone or pterion osicule
    - Anterior fontanelar bone or bregmatic bone
    - Obelisc bone
  - Accessory sutures
    - Diploic and vascular markings
- **Traumatic lesions**
  - Linear fractures
  - Growing fractures
  - Sutural diastasis

**VARIATIONS IN SKULL DENSITY**

- **Generalized decreased density**
  - Bony dysplasias
    - Osteopetrosis
    - Achondrogenesis
  - Metabolic disorders
    - Hypophosphatasia
    - Menkes syndrome
- **Localized decreased density**
  - Normal variants
    - Digital or convoluted markings
  - Lacunar skull
  - Osteomyelitis
  - Generalized increased density
  - Bony dysplasias
    - Osteopetrosis
    - Pyknody sostasis
    - Craniodiaphyseal dysplasia
  - Hemolytic anemias
  - Localized increased density
  - Bony dysplasias
    - Frontometaphyseal dysplasia
    - Craniofrontal dysplasia
  - Rickets

**CALVARIAL MASSES (WITH OR WITHOUT ASSOCIATED LYtic BONE LESIONS)**

- **Normal variants**
  - Prominent external occipital protuberance
- **Vascular lesions**
  - Haemangiomas
  - Venous malformations
  - Sinus pericranii
  - Lymphangioma
- **Congenital inclusion cysts**
- **Cephaloceles**
- **Traumatic lesions**
  - Haematoma
  - Caput succedaneum
  - Cephalocele
  - Subgaleal haemorrhage
  - Fractures
    - Leptomeningal cyst; Infradural arachnoid cyst
  - Infections
    - Post-puffy tumour
  - Neoplasms
    - Benign neoplasms
      - Osteomas
      - Fibrous dysplasia
      - Langerhans cell histiocytosis
    - Malignant neoplasms
      - Melanotic neuroepithelioma
      - Metastatic neuroblastoma
      - Leukaemia

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**Fig. 3:** Semiological classification of cranial vault normal variants and lesions in children.

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**Fig. 4:** Cranial vault deformities due to craniosynostosis: A) Trigonocephaly (metopic synostosis). B) Brachycephaly or bradycephaly (bilateral coronal synostosis). C) Scaphocephaly or dolichocephaly (sagittal synostosis). D) Anterior plagiocephaly (in this case due to a right unilateral coronal synostosis)

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Fig. 5: US: A) Normal metopic suture and B) metopic synostosis.

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Fig. 6: Sagittal synostosis: A) CT superior view 3D and B) axial MIP reconstructions.

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Fig. 7: Unilateral left coronal synostosis. A) CT superior view 3D and B) axial MIP reconstructions.

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Fig. 8: Unilateral left lambdoid synostosis: A) CT posterior view 3D and B) axial MIP reconstructions.

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Fig. 9: Metopic synostosis: A) CT anterior view 3D and B) axial MIP reconstructions.

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Fig. 10: Bilateral coronal synostosis: A) CT frontal view 3D and B) axial MIP reconstructions.

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Fig. 11: Crouzon syndrome. Unilateral left coronal synostosis: A) CT anterior view 3D and B) axial MIP reconstructions.

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Fig. 12: Pfeiffer syndrome. Left coronal and left lambdoid synostosis: A) CT frontal view, B) left lateral view and C) superior view 3D reconstructions and D) axial bone window image.

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**Fig. 13:** Postnatal deformational plagiocephaly. CT superior view 3D reconstruction.

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**Fig. 14:** Skull molding: posterior and superior displacement of parietal bones and elevation of occipital bone. Re-expansion of the cranium after few days.

Fig. 15: Faulty fetal packing: concave depression of the calvarium due to a malpositioned limb in utero (arrow). Plain radiograph.

Fig. 16: Depressed fractures. CT posterior view 3D reconstruction.

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Fig. 17: Small parietal foramina (arrow). CT superior view 3D reconstruction.

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Fig. 18: Abnormally large anterior fontanelle. CT anterior view 3D reconstruction.

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Fig. 19: Large parietal fontanelle. CT postero-superior view 3D reconstruction.

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Fig. 20: Large parietal foramina. A) and B) CT superior and lateral view 3D, C) axial MIP and D) coronal MPR bone windows reconstructions.

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Fig. 21: Delayed closure of sutures in a patient with congenital hypothyroidism. A) CT anterior, B) lateral, C) posterior and D) superior 3D reconstructions.

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Fig. 22: Intrasutural bones. CT posterior view 3D reconstruction.

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Fig. 23: Inca bone (arrow). Antero-posterior plain radiograph.

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Fig. 24: Anterior fontanellar bone or bregmatic bone. A) CT superior view 3D and B) axial MIP reconstructions.

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Fig. 25: Linear skull fracture of the occipital bone. CT posterior view 3D.

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<table>
<thead>
<tr>
<th>Skull fracture</th>
<th>Accessory suture</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lineal sharp lucency</td>
<td>Zigzag pattern</td>
</tr>
<tr>
<td>Non sclerotic edges</td>
<td>Sclerotic edges</td>
</tr>
<tr>
<td>Usually unilateral / Asymmetric if bilateral</td>
<td>Usually bilateral and symmetric</td>
</tr>
<tr>
<td>Can cross adjacent suture lines</td>
<td>Merges with the adjacent suture</td>
</tr>
<tr>
<td>Soft tissue swelling associated</td>
<td>No soft tissue swelling associated</td>
</tr>
<tr>
<td>Possible displacement of its edges</td>
<td>No displacement</td>
</tr>
<tr>
<td>Widens as it approaches a suture</td>
<td>No associated diastasis</td>
</tr>
</tbody>
</table>

**Table 1**: Differential diagnosis between skull fractures and accessory sutures.

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Fig. 26: Mechanism in the formation of a leptomeningeal cyst. A) bone fracture with dural tear and early protrusion of the arachnoid into the fracture. B) Lateral marginal erosion of bone and widening of the fracture. C) Leptomeningeal cyst.

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Fig. 27: Osteomyelitis of the left temporal bone. Axial CT A) Acute stage and B) Healing stage.

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Fig. 28: Prominent external occipital protuberance. Lateral plain radiograph.

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Fig. 29: Frontal congenital inclusion cyst. A) CT anterolateral view 3D reconstruction and B) axial bone window image.

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Fig. 30: A) Cephalhematoma; B) Subgaleal haematoma; C) Caput succedaneum

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**Fig. 31:** Bilateral cephalhematoma. A) CT anterior view 3D and B) coronal bone window MPR reconstructions.

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Fig. 32: Skull eosinophilic granuloma. A) CT lateral view 3D reconstruction and B) axial bone window image.

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Fig. 33: Metastatic neuroblastoma. Post-contrast CT axial soft tissue and bone windows images.

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Imaging findings OR Procedure details

We review paediatric cranial low-dose MDCT scans performed at our centre between 2005 and 2012, compiling all cases with calvarial pathology as well as skull variants that may lead to misdiagnosis.
Conclusion

A wide spectrum of cranial vault congenital and acquired lesions is commonly encountered in children. They frequently show as calvarial deformities, bumps or defects, posing a significant source of concern for parents. Often clinical examination is not enough to reach a diagnosis, therefore imaging studies are crucial for the management of these patients.

Moreover, skull anatomical variants mimicking disease are common imaging findings and radiologists should be aware of them to avoid further diagnostic or therapeutic procedures.
References