

Anterior plagiocephaly – A report of a case and operative technique

Anteriorna plagiocefalija – Prikaz primera in opis kirurške tehnike

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KLjučne besede:

kraniosinostoza;
plagiocefalija;
rekonstrukcija

Key words:

craniosynostosis;
plagiocephaly;
reconstruction

Citirajte kot/Cite as:

Zdrav Vestn. 2017;
86(1–2):34–41

Received: 2. 7. 2016

Accepted: 28. 12. 2016

Abstract

Anterior plagiocephaly is a result of premature unilateral fusion of the coronary suture. It is the third most common form of non-syndromic craniosynostoses. Its main features include forehead, supraorbital arch and anterior cranial fossa deformation as well as orbital asymmetry. The treatment is surgical. The main aim is to relieve compressed brain tissue and achieve an acceptable aesthetic effect. We describe a boy with anterior plagiocephaly, who has undergone an extensive craniofacial reconstruction.

Izveček

Anteriorna plagiocefalija nastane zaradi enostranske kraniosinostoze koronarnega šiva. Je tretja najpogostejša oblika nesindromske kraniosinostoze. Glavne značilnosti so enostranska sploščenost čela in supraorbitalnega loka, deformacija baze sprednje lobanjske kotanje in asimetrija orbit. Zdravljenje je kirurško, saj s tem razbremenimo utesnjeno možgansko tkivo in s popravki dosežemo estetski učinek. Opisujemo klinični primer dečka z anteriorno plagiocefalijo, pri katerem je bila s kirurško tehniko opravljena obsežna kraniofacialna rekonstrukcija.

Introduction

Anterior plagiocephaly is a condition resulting from the unilateral craniosynostosis of the coronary suture (1,2). The clinical picture is characteristic. One side of the forehead and supraorbital arch are flattened with the other half of the forehead compensatory bulged. The changes are most evident in the superior and frontal views. The cranial base is distorted and the orbits are asymmetric. On the affected side, the ear is shifted anteriorly and the temporomandibular joint and nose base may be asymmetric (3-6). Anterior plagiocephaly represents 13 % to 16 % of all craniosynostosis; it is the third most common form of non-syndromic craniosynostoses with the incidence of

1 per 10.000 births. The disease is twice more common in girls, and the right side is usually affected (1,2).

For the diagnosis, the imaging with computed tomography (CT) and three-dimensional (3D) reconstructions are needed for the visualisation of the synostotic suture as well as the complex deformation of the cranium and cranial base. A recognizable feature, visible on radiographs, is the deformation of the ipsilateral orbit, also known as the Harlequin deformation. It is caused by the superior displacement of a large sphenoid wing, giving the orbit on the affected side a characteristic elongated appearance (5,6).

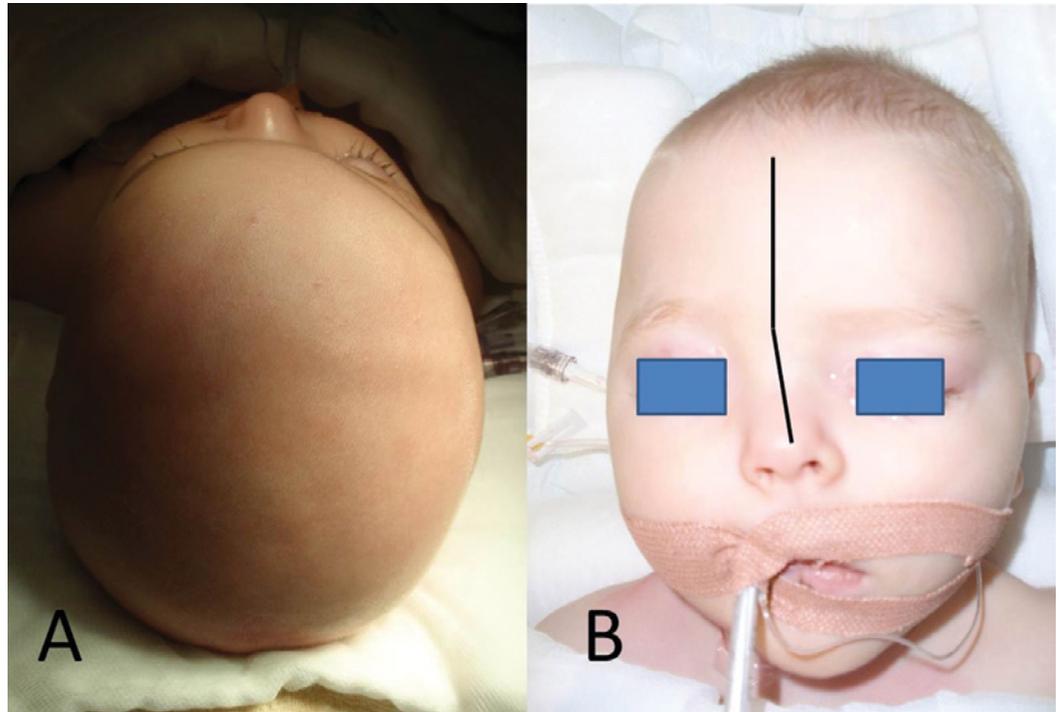


Figure 1: A clinical characteristics of right anterior plagiocephaly with the orbital asymmetry and nose deviation in the frontal view (A). The right side of the forehead and supraorbital arc are flattened and the opposite side of the forehead is compensatory bulged in the bird's perspective (B).

The treatment is surgical with a complex reconstruction of cranial and facial bones. Besides the aesthetic effect, the aim is to prevent the rise in intracranial pressure during brain growth. Up to

30 % of children may exhibit such difficulties (4-7). In this article, we describe a boy with anterior plagiocephaly, who underwent an extensive craniofacial reconstruction.

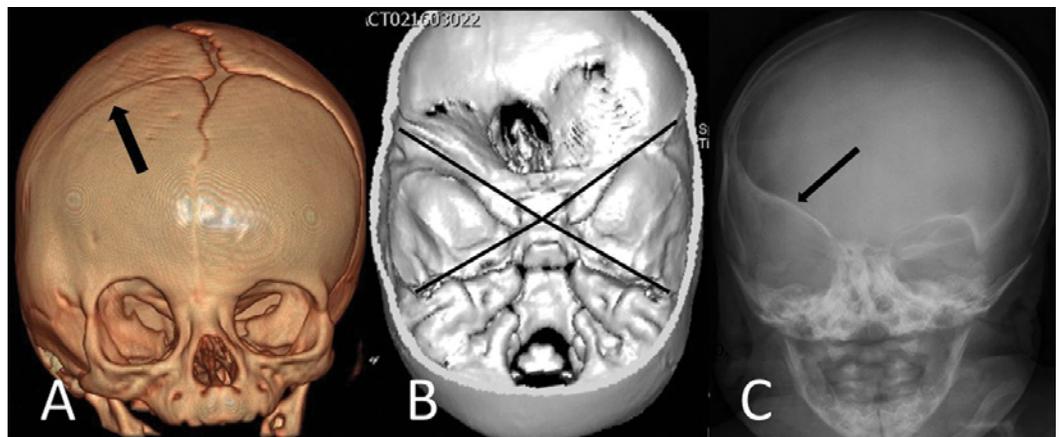


Figure 2: A 3D-reconstruction shows the prematurely fused right coronary suture (arrow) and the deformation of the skull base (B). On X-rays, a typical Harlequin deformity (arrow) caused by superior displacement of the large sphenoid wing can be seen (C).

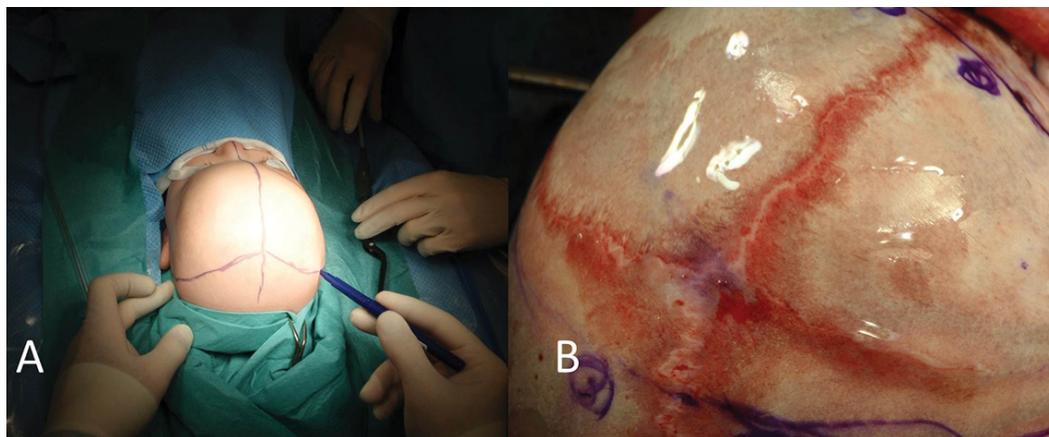


Figure 3: During the operation, the child was in supine position. A coronal skin incision was performed (A). After the periosteal dissection (B), the fused right coronal suture was recognized. The edges of bifrontal craniotomy, running parallel with the supraorbital arch, can be seen.

Case report

A seven-month old boy was referred to the neurosurgical clinic due to unilateral craniosynostosis. On clinical examination, the deformation of the head was evident. The nasal root deviated to the left, the right ear was shifted anteriorly, and the orbits were asymmetric. The right side of the forehead was flattened, the left was bulging forward (Figure 1A, 1B). These features were consistent with the clinical picture of anterior plagiocephaly. A CT scan with 3D-reconstruction confirmed the premature obliteration of the right coronal suture, resulting in the deformation of the head

(Figure 2A, 2B). In order to relieve the cranial distortion and decompress the growing brain, reconstructive surgery was performed. It was performed at the age of seven months. The skin incision was running in the coronal plane (Figure 3A). The skin flap was raised. The periosteum was cut from the left to the right pterion and separated from the bone. Then, the aponeurosis of the temporal muscle was dissected off the sphenoid and temporal bones (Figure 3B). A rise of bifrontal bone flap followed. The craniotomy ran bilaterally to the pterion and to both coronal sutures. The dura was epidurally dissected from the base of the anterior fossa and the sphenoid

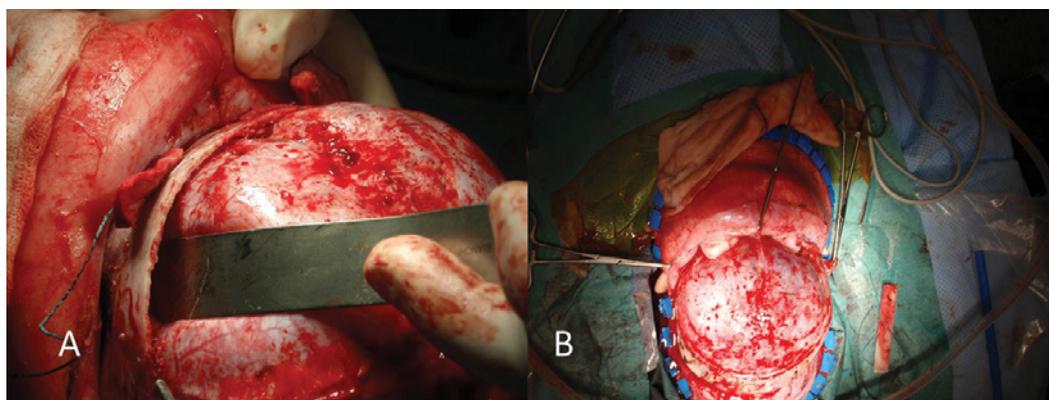
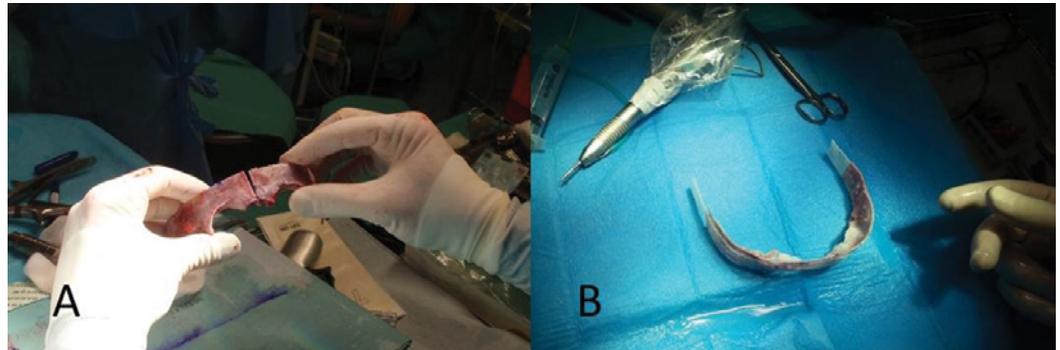


Figure 4: The bone flap was removed and epidural dissection from the anterior fossa followed (A). The *bandeau* was removed and both periorbits were exposed (B).

Figure 5: The supraorbital *bandeau* after the harvesting cut into two halves (A) and after the reconstruction (B).



wings, and then a frontoorbital bone strip (also called a *bandeau*) was removed (Figure 4A and B). The *bandeau* osteotomy was running over the frontonasal suture, both frontozigomatic sutures, and through the orbital roofs. After the *bandeau* harvesting, the remodelling followed (Figure 5A). The *bandeau* was cut in the middle of the supraorbital arches and the asymmetry between the left and the right side was corrected (Figure 5B). The remodelled frontoorbital *bandeau* was finally placed back but in a new position. In this way, the flattened and retracted right orbit was remodelled and the asymmetry was corrected (Figure 6A). The next step included the correction of the flattened frontal bone on

the right side. A new bony forehead was made from the bifrontal bone flap. It was fixed to the frontoorbital *bandeau* with biodegradable osteosynthetic plates. The remaining part of the bone was split and the fragments were used to cover the bone defects (Figure 6B). All fixed fragments were covered by a periosteal flap. Thus, the reconstruction of the facial and cranial parts was completed.

Discussion

Anterior plagiocephaly is a deformation of the skull occurring for unilateral coronal suture synostosis. It causes complex deformities of the face, cranial base and cranial vault. The only possible and

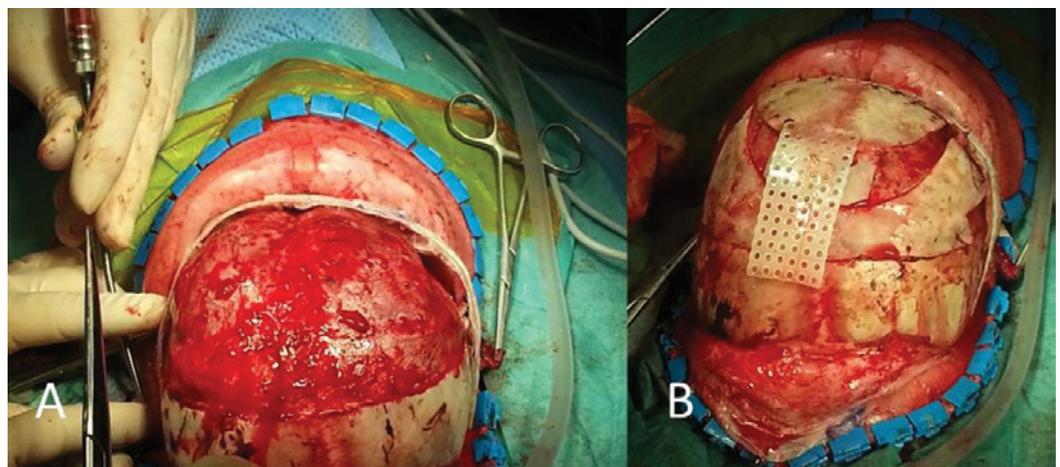


Figure 6: The *bandeau* was fixed to the position with resorbable plates (the so-called *advancement*) (A). The final appearance after the completed reconstruction and filling of bone defects (B).

successful treatment is surgery. Besides having an aesthetic purpose, the surgery mainly aims at correcting the deformation and preventing the intracranial complications, such as intracranial hypertension (7,8).

The surgery for anterior plagiocephaly reconstruction is a complex procedure (7). In anterior plagiocephaly, the growth of the affected side is lagging. In order to minimise the irregularity, the *bandeau* on this side is moved further anteriorly in comparison to the unaffected side, therefore equalising the difference. This advancement provides the symmetry of the supraorbital arc. Recently, many authors have shown a variety of surgical techniques for the treatment of anterior plagiocephaly. In the early 20th century, the so-called strip craniectomies were introduced in the treatment of non-syndromic craniosynostoses. In the sixties, Tessier developed modern surgery for craniosynostoses. The reconstruction of plagiocephaly with hypercorrection is one of the described variations of the basic surgical technique providing better long-term aesthetic results. This means that hypercorrection

is conducted both in the coronary and sagittal planes (9-14). The technique may reduce a possible long-term temporal narrowing of the craniosynostotic side as well as the possibility of synostotic recurrence. Lately, some less invasive methods have been described, such as endoscopic suturectomy (13-16).

The diagnosis of anterior plagiocephaly may be evident as early as in pregnancy on an ultrasound examination. It is important the diagnosis be made in the first months of life and the baby operated within the first 12 months of age. Because of more favourable outcome, the surgery in our patient was postponed to the age of seven months. The optimal time of surgery is still debatable, with most authors recommending surgery between six and 12 months of age (9,10). Early operations may be risky due to a higher recurrence of craniosynostosis and a potential large loss of blood. Late surgery may also be associated with aesthetic and functional complications and increased intracranial pressure. The technique is complex and encompasses a multidisciplinary approach (11,13).

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