

# Positional plagiocephaly: a Task for Primary Care

Guidelines for diagnosis, prevention, treatment,  
monitoring and referral from primary care

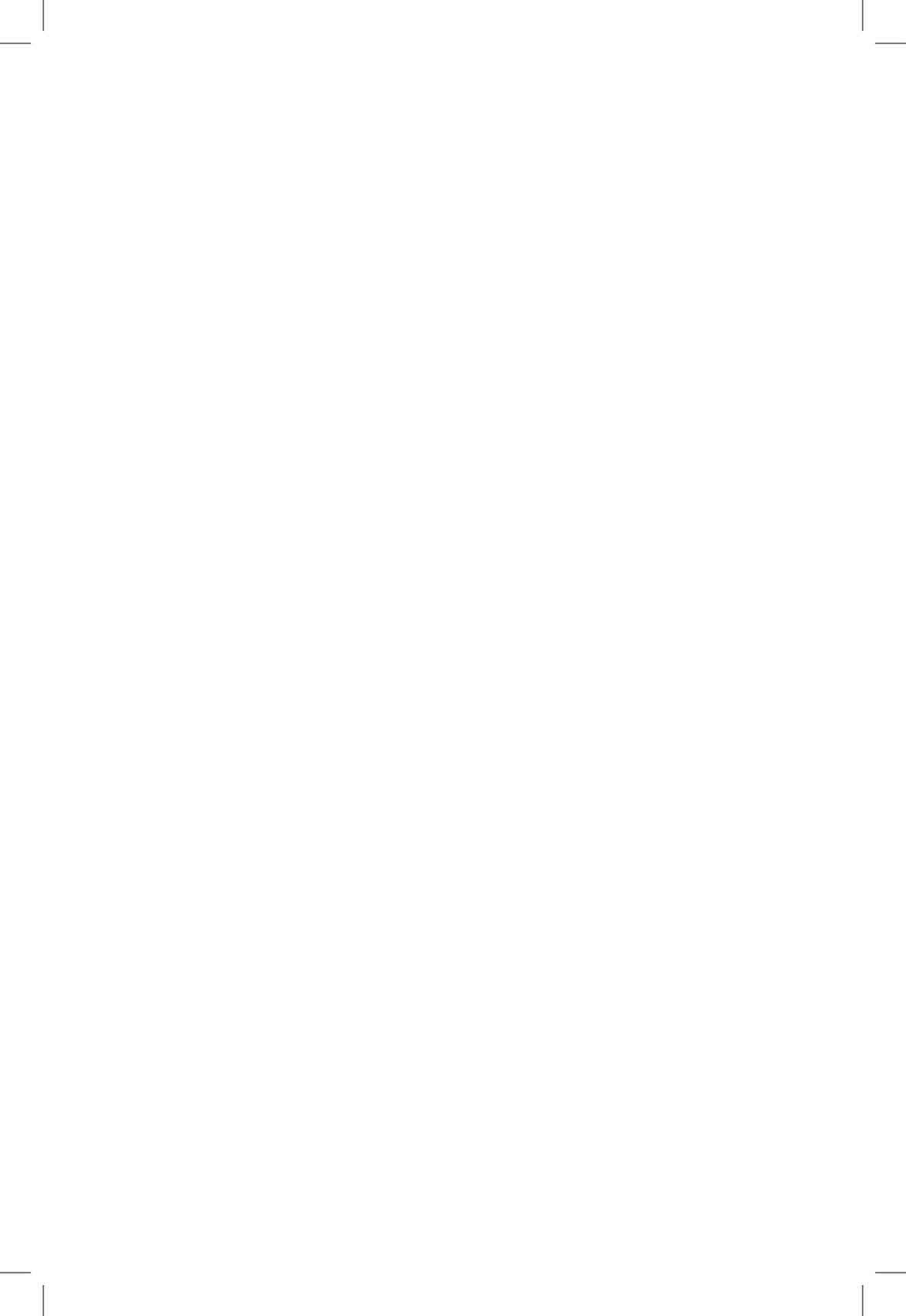
Joan Bosch i Hugas

Primary care paediatrician. EAP Rambla (IG), Sant Feliu de Llobregat

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## Presentation

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Former President of the Spanish Paediatrics Association;  
Honorary Member of the Catalan Society of Paediatrics

I take this opportunity to present the monograph by Doctor Joan Bosch and



Doctor Josep Maria Costa on plagiocephalies to recall that cranio-facial dysmorphism, often found within the large chapter on minor disorders that that one has to know well in practice, has always been

a clinical finding of interest for clinical paediatrics, even though it does not receive due attention in the period of basic medical training and of paediatric specialization.

As with the frequent asymmetry in thigh folds and many other symptoms that it would be long-winded to list, this dysmorphism catches the attention of parents who soon convey their worries to the person in charge of supervising their child. When this happens to the face or cranium, it seems logical for their alarm to increase, faced with the danger of an eventual change in the normal appearance or, even worse, an alteration in the content of the cranial vault: the central nervous system. A check-up is often enough to reassure parents, since usually a defect within the limits of normal asymmetry is found that will be attenuated over time, as so often observed in cranial deformity due to the foetus passing through the birth canal.

Given this, for a long time priority was given to confirming the dimensions of the skull to rule out the feared microcephaly and macrocephaly. For this reason, and also as a means of growth control, newborns' head circumference must still be measured and also in the following check-ups during the paediatric age. In the past it was not uncommon, apart from in relationship with the nervous system, to see generally obvious cranial anomalies such as brachycephaly and wide-fontanelle in rickets, the closure of sutures in cases of hypocalcaemia, craniotabes in premature babies, retardation of ossification in congenital hypothyroidism or the thickening of bones in haematological processes with chronic haemolytic anaemia in children.

Nevertheless, among the changes typical in modern paediatrics, there is an increase in consultations in primary care, neuropaediatrics and paediatric neurosurgery, due to manifest cranial deformity or plagiocephaly, in the broad sense of this term. And within this chapter, the aetiopathogenic conditioning factors have changed

in terms of relative frequency. It is not necessary to go into much detail now, as the reader will soon see these below in the excellent monograph by Joan Bosch and Josep Maria Costa. Positional plagiocephaly has now become predominant by far, with increasing incidence, between 8 and 14 per cent of infants, since the introduction of the decubitus supine position to prevent sudden death syndrome in the early months of the child's life. However, until about fifteen years ago, an interest in craniosynostosis dominated, i.e. the premature closure of the sutures that join the different bones that form the cranium and which lead to a special morphology depending on the suture affected. This has a specific technical name: scaphocephaly or dolichocephaly, brachycephaly, trigonocephaly, and turriccephaly and plagiocephaly, which etymologically means oblique skull.

When these cases were studied, within the large group known as rare diseases, some were idiopathic or of uncertain causes, some were attributed to compression within the womb and others had a genetic basis. More interest was given to the type associated with other anomalies that constituted a syndrome. Now we know that there is often an alteration in one of the receptors of the different fibroblast growth factors, as in the acrocephaly/syndactyly Apert syndrome, the Crouzon craniofacial dysostosis syndrome, the Saethre-Chotzen and Muencke syndromes and the Pfeiffer syndrome involving brachycephaly and broad thumbs. Other well-known craniosynostosis syndromes are the ones found in the corresponding symptoms first described by Bailen-Gerod, Carpenter, Jackson-Weiss and Shprintzen. Clinical recognition of these symptoms is still very important in order to refer the child as soon as possible to neurosurgery. This will lead to full diagnosis and appropriate treatment in order to prevent not only the progression of the deformity, but also possible complications such as papilloedema and optic atrophy.

The last mention in these lines of introduction that I am pleased to be writing is of a more personal nature: in 1998 the Editorial Espaxs publishing house published a work, now sold out, called the Atlas of Paediatric Syndromes (*l'Atlas de Síndromes pediátricos*) in which for my part I was responsible for describing a selection of the symptoms and the summary of what was then known about their clinical relevance,

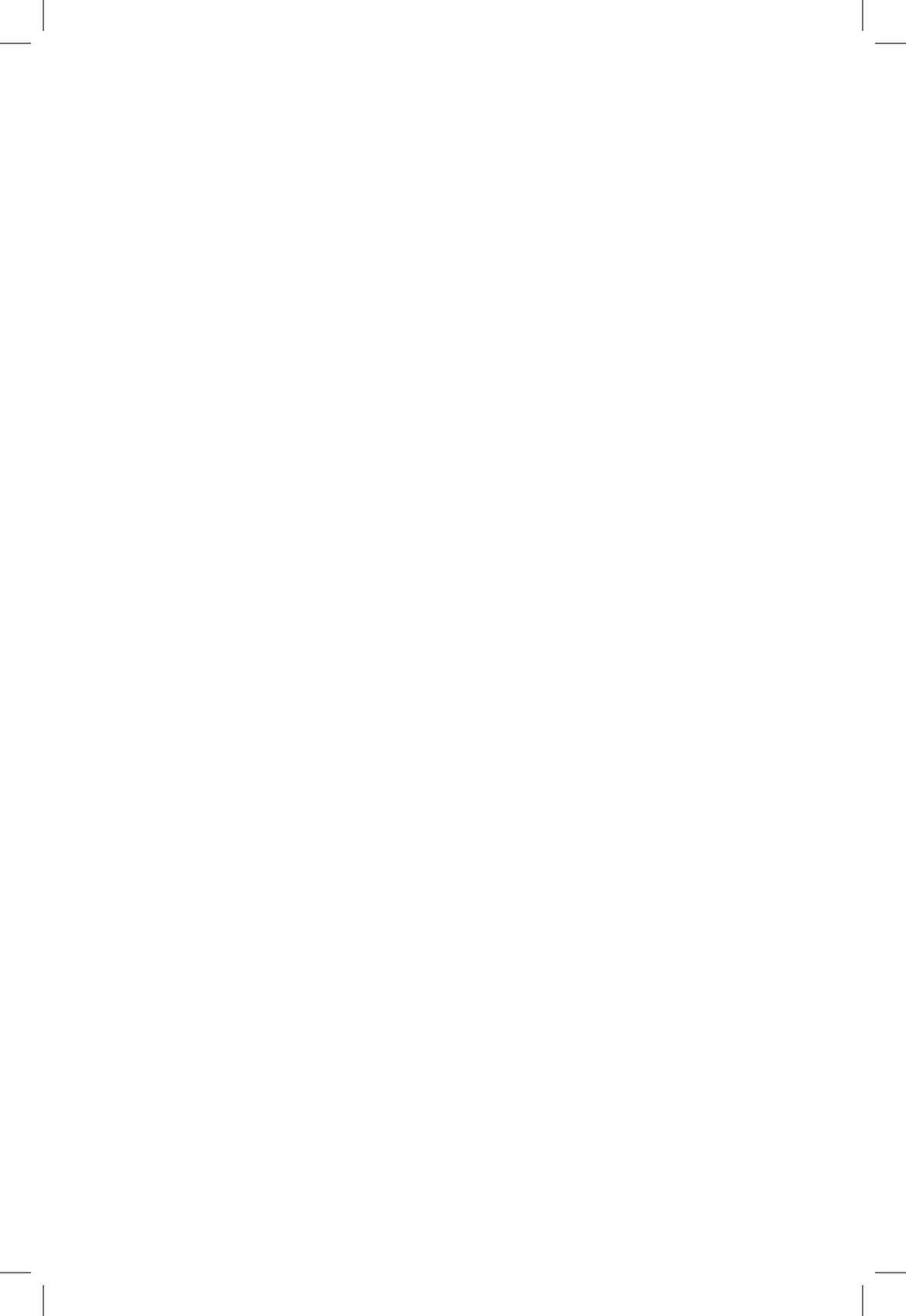
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diagnoses and treatment. But most of the credit goes to Dr. Joan Bosch i Hugas, who skilfully and patiently transferred the material on the slides with his particular drawing technique. He not only achieved a striking resemblance to the models, but highlighted the fundamentals for diagnosis, always guided by his explanation of the pathogenesis. ⇐

In that book, 20 pages were devoted to the matter being presented here, but limited to the syndrome types of craniosynostosis. Dr. Bosch, who started his brilliant career as a paediatrician with me at the Professional School of the Hospital Clinic of Barcelona, had already made some useful diagrams to show in the classroom and in the Manual or Treatise on Paediatrics, which I direct.

It is therefore no surprise that in this book on positional plagiocephaly, in addition to showing his experience, his critical nature and being responsible for the fundamental part of the text, supported by the authority of the children's neurosurgeon J.M.Costa, Joan Bosch helps the reader understand the matter with these excellent drawings. Thus, the concept of positional plagiocephaly, the pathogenic mechanism, the necessary differential diagnosis, the useful prevention or choice of positional, physiotherapeutic or cranial orthosis treatment (cushioning against the skull) are all easy to understand for the main target audience — primary care paediatricians. Moreover, this beautiful monograph will be equally useful for anyone interested in a topic that is very prevalent today, whether they be doctors, nurses, neurosurgeons, health care managers, physiotherapists or the parents themselves. For my part, I am pleased to be among the first to warmly congratulate the authors for their work and to acknowledge the support of all those who make it possible for it to be widely read.





## Introduction

There has been a profusion of papers published in different countries defending the effectiveness of the prevention of Sudden Infant Death Syndrome (SIDS), mentioning so-called “new risk factors”. These mainly include the prone decubitus position for sleeping, excessive covering or atmospheric heat and maternal smoking during pregnancy and after delivery (1-3), which encouraged the international medical community to adopt preventive measures. There was no room for doubt. The studies covered seven countries on four continents, including different races. On avoiding the prone position for sleeping infants, a 50% decrease in mortality for SIDS was predicted. As a result, many countries began campaigns recommending that infants sleep in the decubitus supine position, but they also considered the lateral decubitus position to be acceptable. The prevention campaign “Back to Sleep” was launched in the United States of America, indicating the idea of babies sleeping on their back, which was certainly the campaign with the most media coverage (3). Shortly after introducing the new measures into society, observations began to proliferate in medical journals mentioning morphological alterations of the cranium attributable to changes in positional habits (4-6). Less than a decade after that recommendation, it was estimated that mortality due to sudden death had been reduced by 40% in countries where the recommendations had been implemented (2). The measure was unquestionable, but required adjustments to make it possible to apply it without causing deformation of the cranium. Thus, in 2000 a new campaign began, sponsored by the American Academy of Paediatrics (AAP): “Back to Sleep - Tummy Time to Play” (7), a slogan that promoted a series of resources to free infants’ craniums from the effects of prolonged gravitational pressure against a flat surface.

The figures mentioned regarding the incidence of positional plagiocephaly range between 8% and 14% in infants of under 6 months (5, 8-11), though this was affected by the lack of uniformity in the criteria for selecting the anomalies. These percentages match the data for persistence at adult age (12). On including slight deformities in prospectively planned studies, the prevalence increases to 33% (13). However, it seems that including slight deformities may create alarm that detracts from the true significance of the problem by considering deformations of aesthetically minimal or imperceptible effects in the counts. For primary care paediatricians, modifying this criterion could be the first step towards solving the problem: simply by

considering that slight deformations do not exist, and that there are severe forms detected very early on, we will have started to tackle the root of the problem.

Spain was not indifferent to the prevention of SIDS, and in 1993 the first preventive initiatives began in the Basque Country and Navarre, but it was not until October 2000 when the Spanish Paediatrics Association (AEP in Spanish) began a governmental prevention campaign (2) in collaboration with the Ministry of Health. As a consequence, positional plagiocephaly soon became the most common reason for consultations made to paediatric neurosurgery, as documented in numerous publications in medical literature in Spain (14–18), the European Community (19), Switzerland (20) and the USA (21,22). These figures on prevalence completely match the data obtained by the Neurosurgery Service at the Sant Joan de Déu Hospital in a review of 804 cases of cranial deformities seen from 1996 to 2005 (fig. 1, fig. 2) prompted by the gradual and steady increase in the number of consultations for this reason. However, this increase in positional deformities, which are not really a disease, leads to cases of deformities due to craniostenosis (which are indeed an inevitable primary malformation requiring surgery) being confused with these and referred late for treatment.

An example of this fact is that the number of publications indexed in Medline with plagiocephaly as a keyword multiplied by six compared to those obtained before 1992. The number of pages in Google searches for parent support groups, for companies providing cranial orthoses,

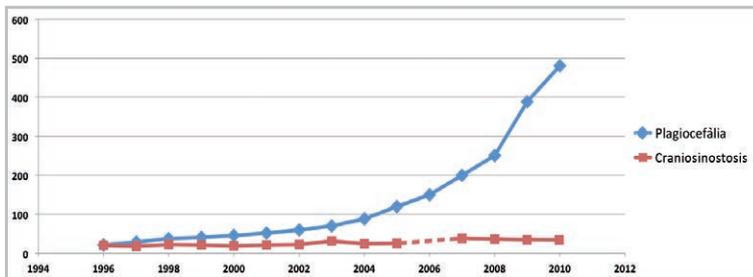


Fig. 1 – Evolution of the prevalence of craniostenosis and positional plagiocephalies from 1996 to 2010 at the Neurosurgery Service of the Sant Joan de Déu Hospital.

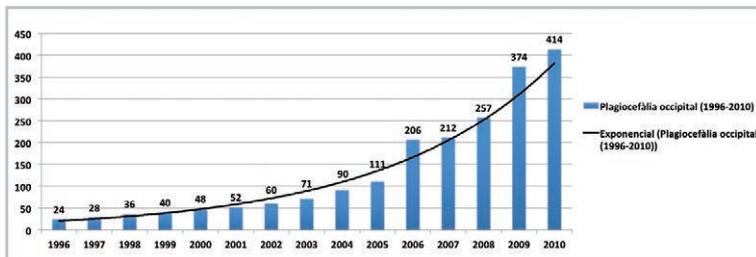


Fig. 2 – Distribution of the prevalence of the 804 cases of positional plagiocephaly attended to at the Sant Joan de Déu Hospital from 1996 to 2005.

for hospital protocols, advice and manufacturers of products related to the preventive positions for infants has also increased exponentially, plunging families into an often tendentious confusion. This means that measures should be taken urgently. The lack of agreed therapeutic criteria gives rise to anxieties and complaints from parents who cannot find a suitable answer to their concerns.

In an attempt to clarify guidelines and treatment, a Consensus-Seeking Day was held in 2006 at the Sant Joan de Déu Hospital in order to channel the problem. This was represented from the medical point of view, inviting neurosurgeons, rehabilitation experts and cranial orthosis specialists, and represented from the social point of view by parents' associations, in addition to the administrative point of view by processing data from the Catalan Dept. of Health to get to know the problem better. All the possible solutions were brought together.

As for the effectiveness of positional measures and the optimum age for hospital referral depending on each kind of pathology, there was complete consensus. This was not the case for recommending cranial orthoses. As a result of this meeting, in 2008 cranial orthoses were included as an additional service to the Catalan health care system, thereby contributing to excessive administrative health care spending. This led us to reconsider what was being done in other autonomous regions and other countries in the European Community. The results varied

greatly as regards the costs of orthoses, ranging from 800 euros in France to 1,500 in Switzerland, and in Spain from 1,200 to 4,000. There was also great variation in terms of the financing system, which in Spain used private health care insurance organisations and subsidies in some regions like Madrid, Valencia or Catalonia.

Nevertheless, the meeting reached agreement on some basic therapeutic criteria to act as guidelines for paediatricians and parents, and on launching a series of outreach sessions at local hospitals and at the Catalan Society of Paediatrics, which pioneered the current proposal.

### **What has happened meanwhile to our field on a tertiary level?**

Table 1 - Number of consultations made in the Neurosurgery Service at Sant Joan de Déu for plagiocephaly, indicating the percentage of orthoses prescribed.

2003 y 2005	120 cases assessed / year	(6% orthoses)
2006 y 2008	200 cases assessed / year	(11% orthosis)
2009	388 cases assessed	(15% orthosis)

From 2003 to 2005, the Sant Joan de Déu Hospital assessed 120 cases a year, in 7 of which orthopaedic correction with cranial orthosis was considered advisable (6%). Between 2006 and 2008, the cases assessed had risen to 200 per year and the number of orthoses recommended had increased to 11%. In 2009, the incidence of cases assessed almost doubled (388 cases) and the percentage of children with orthoses rose to 15%. (Table 1). Far from solving the problem, year on year the lack of a national prevention plan created a higher incidence of positional cranial deformities, a greater number of recommendations for orthopaedic correction and an increase in the social demand for cranial orthoses that was not always justified. These figures of prevalence may increase, since the review by the AAP in 2005 warned of an increased risk of SIDS when the lateral position is chosen, advising against any other position than decubitus supine (23).

## Classification of Plagiocephalies

From an etiological point of view, cranial deformities may be due to extrinsic compressive factors or, less often, to idiopathic premature closure of cranial sutures (craniosynostosis). Chronologically, the extrinsic causes can be intrauterine, intrapartum, or during the first months of life.

Although there are several factors that may alter the proper shape of the foetal head through a moulding phenomenon before and during birth (macrosomia, macrocephaly, multiple pregnancy, uterine myomas, bicornuate uterus, instrumentation of the birth etc.), generally these are resolved within the first six weeks of life (24–26). However, they must be considered as potentially leading to the infant tending to adopt a comfortable position whereby they tend to rest the head on the more flattened side, exacerbating the existing asymmetry and developing a true positional plagiocephaly that cannot then be corrected. This process of mixed causality is particularly evident when there is torticollis, an association that occurs frequently.

To increase the confusion, and possibly leading to delays in taking measures, the plagiocephalies that develop after birth that we are identifying as positional plagiocephalies have been given various names such as: posterior plagiocephaly, occipital plagiocephaly, plagiocephaly without synostosis, postural plagiocephaly, benign positional moulding and deformational plagiocephaly.

## Differential diagnosis of positional plagiocephaly

The cranial shape with plagiocephaly becomes established based on deforming patterns that vary in intensity but which are constant, enabling them to be identified. Infants placed in the decubitus supine position tend to move the head to one side, rarely managing to rest the cheek, with rotation limited to about 45 degrees. In this position, the constant pressure from gravity on the parieto-occipital region on one side causes flattening of the affected area, moving the petrous portion of the temporal forward and down and causing compensatory bossing of the contralateral occipitoparietal region. The result is a parallelogram distortion which, due to the rotation of the cranial base that resists deformation by crushing, causes anterior displacement of the entire affected side, dragging the ear with it, and may cause ipsilateral frontal bossing or, depending on the severity, may even affect the facial morphology on the same side while the contralateral area is displaced backwards. The resulting facial appearance is asymmetric due to a greater opening of the palpebral fissure and greater prominence of the orbital rim and the cheekbone. The distance between the tragus and exocanthion of the eye is shortened and the ear often stands out from the skull abnormally. The nasal root is usually centred. (Fig.3, fig.4). It does not seem too far-fetched to assume that when the term plagiocephaly was chosen, "oblique head" in Greek, it referred to positional plagiocephaly, even it was to end up meaning all asymmetric deformations of the cranium.

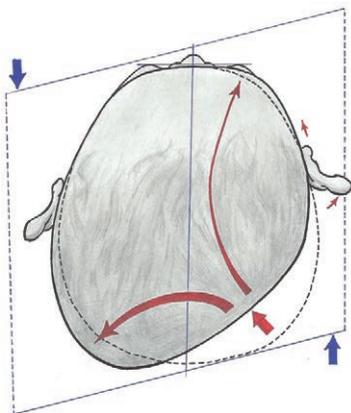


Fig. 3 – Diagram of the pathophysiology of positional plagiocephaly.



Fig. 4 – Overhead image of a positional plagiocephaly.

However, when the affection is bilateral due to continuous pressure on the occiput without a preference for one side, the cranium takes on a more or less symmetrical shape, also known as pachycephaly, in which encephalous growth puts pressure on the parietal areas, causing an increase in the cranium's biparietal diameter and the vertical, which increases up to the lambdoid sutures. Despite this, it does not have a significant effect on facial morphology and may be undetectable from a frontal view. In the most severe cases, the very characteristic profile gives the impression that the back of the skull has been cut off, lining itself up with the neck without showing any curvature (Fig. 5.1, fig. 5.2, fig. 6).



Figura 5 (1 -2).

Fig. 5.1 – In the frontal view, the increase in interparietal distance is not enough to confirm a brachycephalic cranium.

Fig. 5.2 - The profile of the brachycephaly gives the impression that the occiput has been cut off.

Orthopaedic correction of positional brachycephalic deformation is more difficult than that of positional plagiocephalies because the posterior flattening of the base of the cranium is less receptive to correction.

Cranial orthoses can modify the shape of the cranial vault, but have little effectiveness in correcting deformities of the cranial base, so that priority must be given to hospital referral.



Fig. 6 – In the overhead view of brachycephaly, a shortening of the antero-posterior diameter is seen, and the increased biparietal distance.

Differential diagnosis must be carried out with craneostenotic occipital plagiocephaly, a very infrequent affliction, even among different types of plagiocephaly due to craneostenosis. The incidence of plagiocephaly due to early closure of cranial sutures, considering all the possible forms it may take, is estimated at 1 per 1,000–2,000 newborns, with data putting lambdoid craneostenotic plagiocephaly at 1 per 33,000 births (27). In other words, the attitude of a primary care paediatrician faced with a posterior plagiocephaly should be to consider it to be positional plagiocephaly until proven otherwise, since after an entire career spent in paediatrics the vast majority have never encountered a lambdoid craneostenotic plagiocephaly.

The shape of a cranium affected by lambdoid synostosis differs from the rhomboid shape characteristic of positional plagiocephaly by taking on a trapezoidal appearance. When the lambdoid suture closes too soon, cranial growth is impossible in its area of influence, forcing the encephalous mass to compensate for the lack of space by bossing the contralateral parietooccipital region, usually more parietal than occipital, due to the greater thickness of the occipital area. As a result, the petrous part of the temporal bone is pushed back behind the ear. Unlike positional deformation, the frontal part also bosses, but contralaterally to the affected side, with an uneven flattening becoming more apparent ipsilateral to the abnormal suture. (fig. 7, fig. 8, fig. 9)

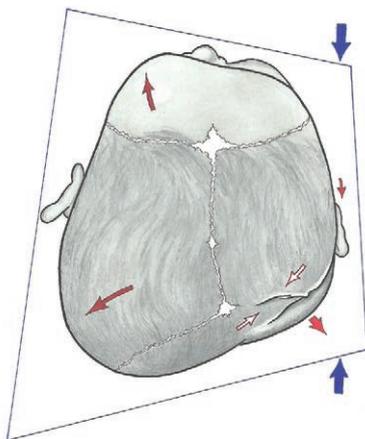


Fig. 7 - Diagram of the pathophysiology of lambdoid craneostenotic plagiocephaly.



Fig. 8 - Cranium affected by craniosynostosis due to premature closure of the left lambdoid suture. Note the irregularities in the flattest area, not observable in positional plagiocephaly.

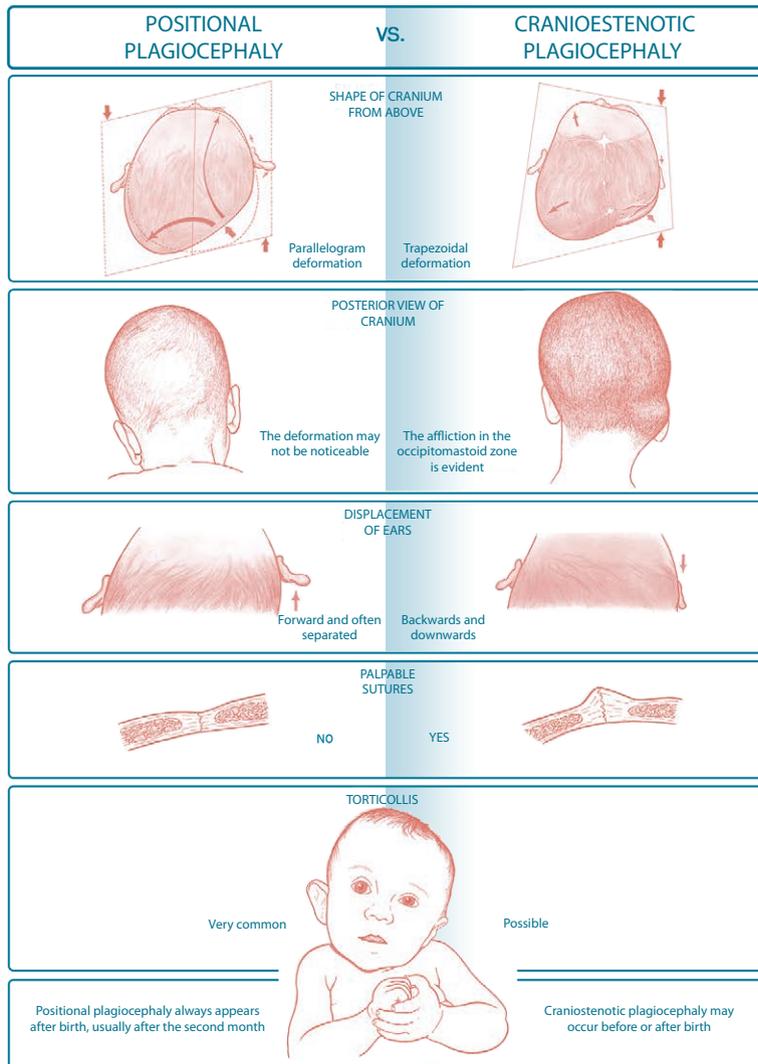


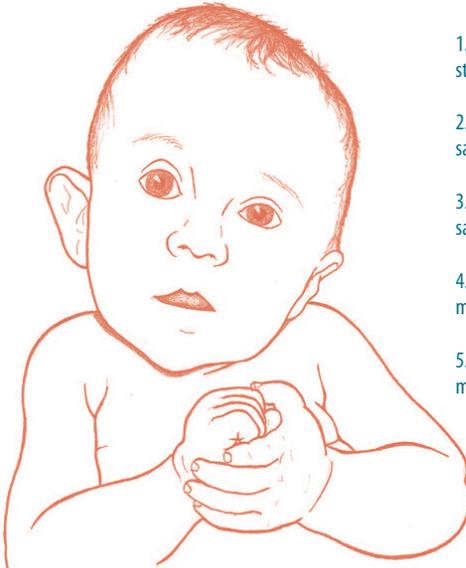
Fig. 9 -Differential diagnosis between positional plagiocephaly and lambdoid craniostenotic plagiocephaly.

On those occasions when the craniostenosis is part of a polymalformative syndrome (Apert syndrome, Saethre-Chotzen syndrome, Crouzon's craniofacial dysostosis etc.), the diagnosis is practically visual with no possibility of confusion with positional plagiocephaly.

## Examination method

The study of positional plagiocephaly should never be limited exclusively to an examination of the head. It is essential to carry out an orthopaedic examination, focusing on the mobility of the neck and the posture of the spine.

Craniofacial examination must be thorough; not limited to what is most obvious. One should check for asymmetries in the face, observing it from the front and in profile: does one side of the forehead stand out more than the other? Are the palpebral fissures the same size? Are the superciliary arches located at the same height? Do the zygomatic arches protrude to the same extent? All of this information indicates the degree to which the deformation has progressed, since it shows the steps in a set pattern of deformation. However, the sometimes striking “detachment” of the ear may be present or not, regardless of the severity. (Fig.10).



1. Does one side of the forehead stand out more than the other?
2. Do the palpebral fissures have same dimensions?
3. Are the superciliary arches at the same height?
4. Does one orbital border protrude more than another?
5. Does one zygomatic arch protrude more

Fig. 10 – Deformidades craneofaciales.

On inspecting the cranium, it is most necessary to examine it from above, as this enables us to observe the changes that may have occurred to alter the ovoid cranial shape and provide answers to new questions. Are the auricular pavilions at the same height? Are the distances from

tragus to endocanthion of the eye the same? The state of the sutures should be assessed with particular care. Faced with a protruding, palpable suture, one should suspect craniosynostosis, whereas palpation of a soft suture is a sign of good prognosis for positional plagiocephaly. With premature closure of sutures, mostly before birth, the overlapping of the sutures' edges that is frequently observed in newborns should be carefully assessed in order to avoid falsely diagnosing a craniostenotic crest. In the overlap, slight pressure on the suture will reveal the lack of closure.

Between 25% and 75% of positional plagiocephalies occur with some degree of limitation to movement of the neck (28-29). Together with congenital muscular torticollis, where palpation reveals intramuscular tumours or one sees fibrotic shortening of the sternocleidomastoid, others also coexist that are more frequent, in which muscular examination is normal, where neither thickening nor tightness is detected but lateral twisting still persists. So, the debate remains open as to whether the deformation is the cause or consequence of laterality. Whatever the initial cause, it is clear that a feedback loop is being established that limits the movement of the neck and thus causes the infant to choose a comfortable position by invariably adopting a position of rest on the same side, which will affect the neck's mobility and in turn increase the deformity. Cheng et al. reviewed 1,086 cases of congenital muscular torticollis and found different degrees of facial symmetry in 90 % (30).

Examination of neck muscle dysfunctions should include a targeted anamnesis. In small infants showing a preference for one breast, leaving one quickly to feed on the other, this should be seen as a possible sign of discomfort due to torticollis. Systematically carrying out chin—shoulder and ear—shoulder stretching exercises for the neck, described below, enables resistance to rotation of the neck and lateralization of the head to be identified. The “spinning stool test”, in addition to reinforcing the diagnosis, may be useful in encouraging parents to follow the necessary physiotherapeutic measures that should be taken. The procedure is simple. The examiner sits on a rotatable stool opposite the parents and holds the infant sitting on his/her lap. The parents attract the infant's attention so that he/she looks at them. The examiner turns to one side and then to the other, noting whether the child is able to keep looking at them by turning his/her head or if he/she turns the whole body or loses eye contact. The main limitation as a tool for early diagnosis arises from the need for the child to acquire appropriate

cephalic control, a circumstance that limits its use to children older than 3 months.

If one considers how often the primary care paediatrician discovers a problem with a hip joint, for example, and no one disputes the need to perform Ortolani and Barlow manoeuvres systematically, it does not seem to make sense not to include stretching exercises as a habitual means of diagnosis in the early months of life, considering the data on the prevalence of torticollis.

Examination of the dorsal and lumbar spine in the prone decubitus position is justified in order to possibly detect scoliotic postures that can be observed relatively frequently.

In the absence of any evidence of a cause–effect association between positional cranial deformities and signs of cerebral dysfunction such as psychomotor retardation, hearing or language problems, visual defects or ADHD, some authors' insistence on singling out neurological examination in these cases does not seem justifiable and is often affected by conflicts of interest. The articles published touching upon this issue simply highlight the concomitance and do not provide information to indicate that correction of the deformity resolves or changes the associated neurological problems (43,14,15), even though there is no doubt that neurological disorders accompanied by hypotonia and decreased physical activity are associated with positional alterations in the cranium far more often than in the healthy population.

Even recommendation of surgery in most craniosynostotic plagiocephalies must be seen as an aesthetic question, since only the most severe examples will affect the eyes, hearing, facial mass or endotracheal hypertension.

X-rays in primary care are not justified as they do not provide any additional data to the physical examination. They could be useful to confirm or discard craniosynostosis, but the possible presence of intracranial bony bridges, local fusions or stenosis and sclerosis of the sutural edges can lead to confusion that make it non-advisable to carry them out systematically (Fig. 11)

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Fig. 11 - X-ray image of a positional plagiocephaly in which one can see images of sclerosis on the lambdoid sutural edges that may be confused with craniostenotic pathology.

The techniques to be chosen for differential diagnosis when in doubt, from the hospital area, are the cranial CT-scan and three-dimensional reconstruction (19).

## Anthropometric determinations

Anthropometric measurements are useful for quantifying the severity of deformities and for objectively observing their progress. To make them, one must have a big enough cephalometer, craniometer or calliper to go around the head of the infant. The easiest tool to handle is Infocefalia's craniometer (Fig. 12), which improves on the cephalometer devised by Bertillon (Fig. 13) due to its smaller size and ability to convert the circumference fractions determined by the Bertillon into linear measurements that can be superimposed on the determinations on the typical photos common to neurosurgeons. With this cephalometer, one can get the Cephalometric Index for Plagiocephaly which, together with the now standardized determination of the cranial perimeter, make up the three necessary measurements to properly evaluate and monitor deformities.



Fig. 12 – Infocefalia's craniometer. Designed by the engineer David Verde, this has significant differences from the classic Bertillon cephalometers. This is not only due to its size, easy handling and much cheaper price, but because it applies mathematical concepts to correct the measurement of fragments of circumference, as the Bertillon does, converting them into linear measurements to compare them to those obtained on photos (a common practice in neurosurgery), making it indispensable for primary care monitoring.



Fig. 13 – Bertillon cephalometer. Devised by Alphonse Bertillon (Paris, 1853 - Switzerland, 1914) a French criminologist who encouraged

The **Cephalometric Index** (IC) enables the degree of elongation (dolichocephaly or scaphocephaly) or flattening (brachycephaly) of the cranium to be assessed numerically. It is calculated by multiplying by 100 the result of the ratio between the maximum biparietal distance and the anteroposterior distance taken at the midline (fig. 14). The value for ideal proportions has been determined as 80. Any value lower than this quantifies the severity of dolichocephaly, whereas all higher values give the degree of brachycephaly, with all values between 75 and 85 ( $80 \pm 5$ ) being considered normal.

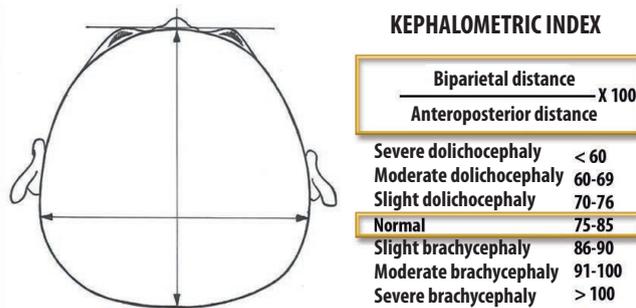


Fig. 14 - Calculation of the Cephalometric Index.

Based on the Cephalometric Index, one can classify deformities into slight, moderate and severe:

Brachycephaly: slight (CI = 80-90), moderate (CI = 91-100), severe (IC > 100)

Dolichocephaly: slight (CI = 70-80), moderate (CI = 60-69), severe (IC < 60)

To determine whether a cranium is plagiocephalic, one does exactly the same thing, i.e. the anteroposterior distance is measured in the mid-line drawn through the centre of the nose perpendicular to the line joining the cheekbones. (Fig.15)

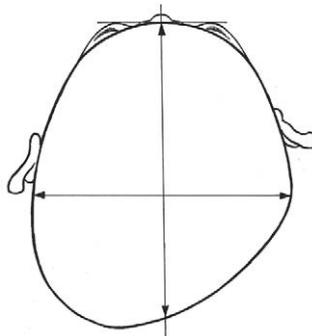


Fig. 15 – Calculation of the Cephalometric Index for a plagiocephalic cranium.

The **Plagiocephaly Index (PI)** or Cranial Asymmetry Index gives the degree of asymmetry between the long and short diagonals of the cranial ovoid (fig. 16). This is considered to be a slight deformity if the value is less than 10 mm, moderate between 10 and 20 mm and severe over 20 mm.

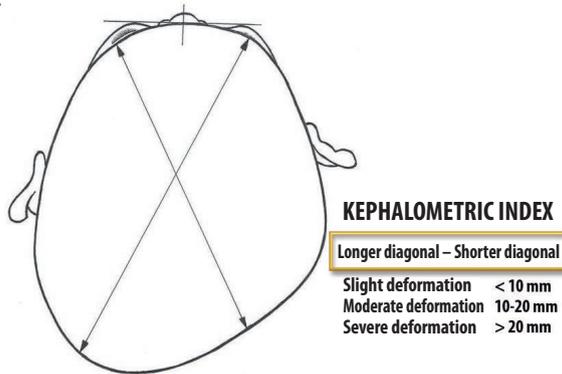


Fig. 16 - Calculation of the Plagiocephaly Index or Cranial Asymmetry.

There is no strict, unanimous criterion as regards determining the diagonals. For Esparza et al., in keeping with other authors (20), there are three valid methods. According to the first, the longest diagonal corresponds to the distance between the external edge of the orbit ipsilateral to the flattening and the most affected point of the bossed occipital region, and the shorter one is the distance between the external edge of the orbit contralateral to the flattening and the most affected point of the occipital flattening. The second one is of little use in primary care, since it would require taking photographs, calculating the two diagonals by drawing lines at 30 degrees to the left and right of the anteroposterior midline. Nor does the third option seem of practical use for paediatric primary care, since it determines the frontozygomatic junctions and the occipitoparietal joints. The Department of Neurosurgery at the Sant Joan de Déu Hospital chooses to recommend referral to the second option as the most practical method for determination in primary care. This involves applying an elastic band, placed as though measuring the cranial perimeter, upon which two parallel marks have been made that are equidistant to anterior and posterior marks that are centred on the anteroposterior midline; i.e. with the nose and the external occipital protuberance acting as reference points for reference points. The crosses determine the points for applying the craniometer, making the determinations uniform without the need to take photographs (fig. 17, fig. 18). Other

determinations, such as the distances between the tragus and the exocanthion of the eye, while no doubt useful, are not essential from the perspective of primary care in order to carry out monitoring and decide on possible referrals to neurosurgery.

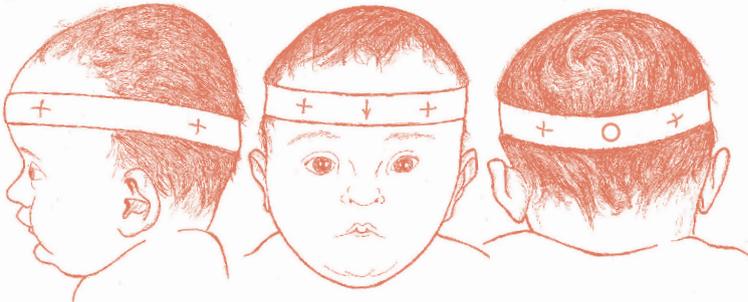


Fig. 17 – The homogenizing band sets points that are equidistant from the centre of the forehead and from the occiput, equallina measurements taken in the transverse plane.



Fig. 18 – Taking measurements with inforcealia's.

Racial or family characteristics may change the “normal” default values for dolichocephalies and brachycephalies, but not for plagiocephalies, which will always be deformational. A slight form of brachycephaly or dolichocephaly, or even a moderate one, may correspond to a family or ethnic trait without, of course, any pathological significance. The increase in Chinese population in our society, for example, has led us to conclude that the normal benchmarks for Caucasians are not the same as for this ethnic group, who tend to have more brachycephalic values as their norm.

The anthropometric study at a tertiary level benefits from techniques that enable three-dimensional reconstructions of the cranial structures. The Department of Neurosurgery at the Sant Joan de Déu Hospital has a mechanism — the Star TM scanner (fig. 19), which enables these assessments to be carried out in three dimensions, thereby providing accurate measurements and avoiding having to make plaster moulds to make ortheses, and providing a fast, simple and non-invasive way of monitoring and evaluating the therapy..

To carry out the test, it is necessary to put a tubular elastic mesh on the child as if it were a compressive cap, which minimizes distortion from the hair when taking cranial measurements (fig. 20.1). The child is placed in the machine in the decubitus supine position (Fig. 20.2) and in a few seconds a system of lasers coupled with computer software generates a three-dimensional reconstruction (fig. 20.3, fig. 21). This gives the values that the same software converts into graphics that can be superimposed on different determinations to see the progress of the deformation. The determinations [results] can be in two dimensions, thereby allowing one to select the cross-section area (fig. 22) or two-dimensional representations of three-dimensional volumetric determinations (fig. 23)



Figura 19 – Star scannerTM



Figura 20-1 – An elastic tublar mesh is put on the head like a cap.



Figura 20-2 – He/she is placed in the machine in decbitous supine.



Figura 20-3 – The computer system obtains the data.



Fig. 21 –Three-dimensional image obtained with Star scanner TM

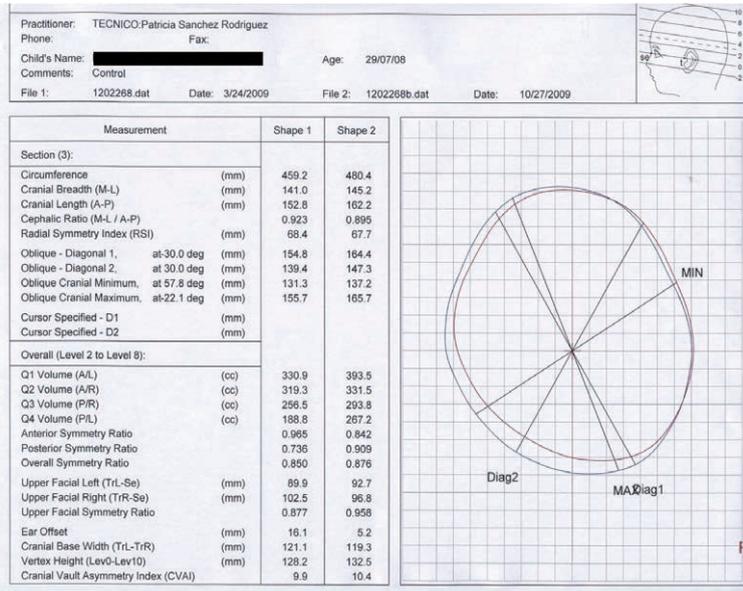


Fig. 22 – Graphical representation of the cranial contour in the chosen cross section associated with the profile seen in the upper right corner. To check on the progress of the deformation, two examinations taken at different times are superimposed. The diagonals drawn determine the measurements necessary to calculate the Cephalometric and Plagiocephaly Indices.

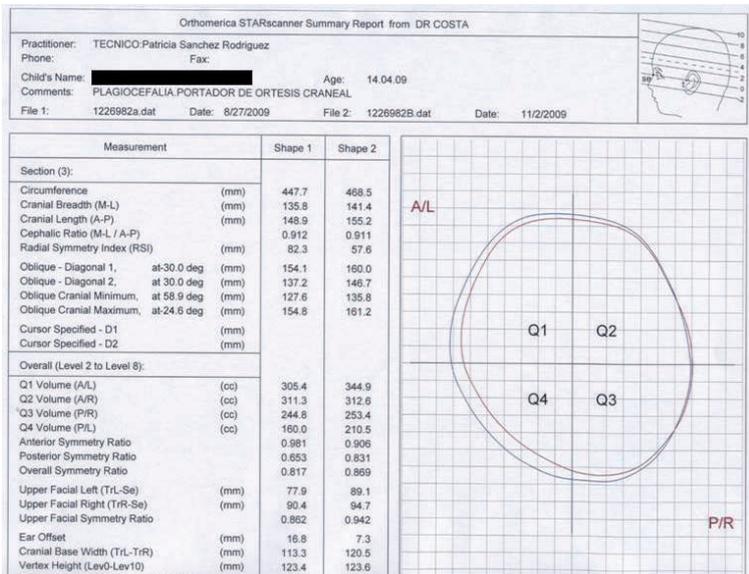


Fig. 23 – Graphical representation of cranial contour by showing two-dimensional representations of three-dimensional volumes of the four areas determined by drawing a sagittal plane through the centre from the nose and a frontal one located on the longest line between the parietals.

## Primary prevention of positional plagiocephaly (table 2)

Table 2 – Prevention of positional plagiocephaly.

1. Systematic practice of stretching exercises for the neck.
2. Always sleep face up. <ul style="list-style-type: none"> <li>- Rotate the head alternately every time the infant sleeps.</li> <li>- Change the child's orientation in the cot every day.</li> </ul>
3. Put the child on his/her belly at times, while he/she is awake and supervised. <ul style="list-style-type: none"> <li>- This must start as of 4 weeks of age.</li> </ul>

Truly effective prevention of positional plagiocephaly can only be considered by taking action as of primary care.

Considering the high degree of concomitance, ruling out torticollis by systematically doing neck-stretching exercises should be accepted as routine.

It is necessary to inform parents and carers — sternly but without causing alarm — about the reasons for the recommended postural advice and the importance of acquiring habits upon which the development of the infant's head may depend. Changes in long-established habits can cause reticence and are not always well received even among health care staff. An unjustified measure can easily be seen as a passing fad or trend of questionable value.

From the first day of life, the sleeping position should be decubitus supine, and the decubitus lateral position (until recently considered safe) is now considered inadvisable (14).

A routine should be established that avoids a fixed positioning of the head, whether this is with the head to one side or staring at the ceiling. Thus, the head position should be rotated alternately to one side and to the other every time we put the infant to sleep during the day and the child's orientation in the cot is to be changed every night. In this way, as the child perceives sounds and movements around them, they will not always turn their head toward the same side.

- As of the first month, when he/she is awake and alert, he/she should sometimes be placed on his/her belly. In this way, the cranium will be freed of the continual pressure of gravity, thereby strengthening the neck and spine muscles more and preparing him/her for crawling. It is very probable that, in the very beginning, he/she doesn't like it and cries, but little by little

he/she will get used to it. It will be a good time to play and bond with the parents and carers.

- To help exercise the head for longer, it is very useful to place a rolled-up towel, a pillow or even one of the parents' or carers' legs between the chest and the floor.
  - As of the third month, placing him/her in front of a mirror and using toys with lights and sounds will encourage the position to be maintained, encouraging them to exercise their senses.
  - At any age, games that avoid the decubitus supine position should be encouraged, placing them in the decubitus prone position on their belly on the parent's knees, making them "fly" like Superman, etc., or any resource the parents can think of that frees the infant's cranium from external pressures while having fun with the parents.
  - In prams, one must prevent the head from remaining on the same side for long periods of time.
  - Baby-carriers may be a good choice. The infant should preferably be placed facing the carrier so that the curve of the spine is not subjected to anti-anatomical positions, and with the knees slightly raised above the buttock area so that they are sitting, but not hanging with most of the pressure on the genital area. In this position, their legs are "open and free" — the ideal posture to prevent and treat hip dysplasia.
  - Pillows for head support designed to reduce cranial pressure can be useful, but we know of no comparative study with positional measurements to demonstrate that it is more or less effective. Until there are studies and evaluations that do not come from the manufacturers themselves, we believe it is advisable to use them when the infant has acquired a degree of independence where it is impossible to maintain suitable positions, but the cranial deformity persists, taking as a base the theoretical effectiveness of the pillows. These may be especially advisable when, for whatever circumstance, the infant does not maintain the preventive positions. Using them in prams could be a good initiative, since the increase in hearing and sight stimuli make it especially difficult to maintain positions. These pillows have a hollow part in the middle that enables them to better adjust to the head, increasing the contact surface and spreading the pressure exerted by gravity. Not all pillows on the market are recommended. The ones made of viscoelastic
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might seem to be the most suitable material as they adapt to the body according to the pressure and heat the latter transmits, but this is a material that does not allow air flow. If an infant turns, he/she could suffocate. An attempt has been made to solve the problem by making holes in the foam, but this does not seem sufficient, since the surface of the foam is still very high in pillows on the market we have been able to study, and they do not ensure safety if the infant buries his/her face in them. Others, filled with polyester fibre, do not even withstand the weight of the head and are totally useless. In fact, we only know of one pillow stuffed with a three-dimensional structure of polyester that enables air flow and keeps its shape, spreading the pressure properly (Mimos<sup>®</sup>).



## Treatment of positional plagiocephaly

(table 3)

Opinion largely agrees that treatment of positional plagiocephaly should be phased: if physiotherapy does not correct the deformity, the use of cranial orthoses techniques will be assessed, and if these are not effective surgery may be used as a last resort, but this should not need to occur. (26-29).

Table 3 – Treatment of positional plagiocephaly.

1. Active positions. <ol style="list-style-type: none"> <li>1. Putting the child to sleep on the bossed side.</li> <li>2. Placing him/her so that he/she has to turn on the bossed side to look at the parents</li> <li>3. Performing stretching exercises for torticollis.           <ul style="list-style-type: none"> <li>- Do these 3 to 5 times a day.</li> <li>- Repeat each exercise 3 to 5 times.</li> <li>- Hold the forced position for about 10 seconds.</li> </ul> </li> </ol>
2. Dedicate some time to the child being face down.
3. Carry out check-ups 7-15 days after diagnosis and then monthly check-ups.

### 1. Physiotherapeutic measures.

The positioning of the head must be active:

- For sleeping, the head must be resting on the bossed side. To achieve this, we can use a rolled up towel, a doll or any resource which, placed as a barrier to prevent turning onto the flat side, makes it possible to hold the posture. If this cannot be done due to the age or temperament of the child, a useful idea is to place an object between the mattress and the cot base, acting as a wedge to raise one side and cause the child's head to move towards the side on which we want them to rest.
- The orientation of the child in the cot must be such that he/she is forced to turn his/her head onto the bossed side when he/she wants to look at the parents.
- Toys are to be offered to him/her always from the bossed side for the same reason.
- When changing nappies, bathing or when giving him/her food, one must always try to position oneself on the side that encourages the desired rotation.

With torticollis, the parents should use manoeuvres for twisting muscles with regular supervision by the paediatrician, who will show them how to do so correctly, paying special attention to boosting their confidence in their own ability to do so correctly. Parents' concerns about the child's protests and their fear of doing him/her harm can only be allayed with proper information about the probable evolution of the deformity left to its own inertia, and by reinforcing their own assuredness in carrying out the manoeuvres.

There are two exercises recommended to properly rotate the neck and lateralize the head:

**Chin-shoulder exercise:** This is designed to restore normal head rotation to the right and left. It is done with the infant in the decubitus supine position. With a hand placed on the child's chest, the shoulders are prevented from coming away from the plane of support, while the other is placed on the side of the face, making the head rotate in an attempt for the chin to touch the shoulder. When the rotation reaches a point of resistance, the position is held gently but firmly for ten seconds. Prolonging this any longer is not usually well received by the parents, who usually relax the pressure to the point that it loses effectiveness. Even if the muscular affliction is on one side, generally on the side opposite to the flattening, we advise doing the exercise on both sides. This will enable the parents to establish a reference for comparison and not to abandon the rehabilitation too soon (fig. 24)



Fig. 24 – Chin-Shoulder exercise.

**Ear-shoulder exercise:** designed to restore lateralization of the head. With the child in decubitus supine position, one hand is placed on the shoulder opposite the side to which we wish to tilt the head, in order to stop it from moving. The other hand, placed on top of the head, tilts it in order for the ear to touch the shoulder. When the point of resistance is reached, the position is held for ten seconds (fig. 25)



Fig. 25 – Ear-Shoulder exercise

Both exercises should be repeated 3 to 5 times a day each, until complete rehabilitation in the neck movement is achieved. A good time to do this may be when changing nappies.

Similarly, as a preventive measure it is advisable to allocate time for games with the child resting on his/her belly.

Following the positional guidelines given, which the paediatricians in primary care are perfectly capable of carrying out, most children are cured and neurosurgery is not necessary, provided that the positional treatment has been applied before the age of 5 months. After this age, the autonomy acquired by the child as regards mobility of the head makes it far more difficult to apply.

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## 2. Cranial orthoses. (fig. 26)

There is little information about the effectiveness of orthoses that is not susceptible to a conflict of interests. They are mostly for commercial or professional purposes with a franchise as regards the product. Recent breakthroughs have created universal access to information, which has provided a soapbox to promote these products without having to be obligatorily screened by scientific journals. The truth is that there are no suitable, planned studies that show greater effectiveness of cranial orthoses as compared to active positioning and muscular stretching (31). In the insufficient comparative studies published there are no significant differences seen after the three months of treatment. (31-33)

Even so, if applied with the correct advice, their efficiency is perfectly proven.



Fig. 26 – Cranial orthoses: an ally that should never be replaced by positional physiotherapy.

It is true that consensus is yet to be found, such as regarding the age when cranial orthoses are to be recommended - 5 months of age - a decision that corresponds to the specialists in paediatric neurosurgery or rehabilitation.

As regards the therapeutic protocol at the tertiary level, the Catalan Institute of Health has approved its use, taking responsibility for the funding, for moderate or severe positional plagiocephalies in children aged between 5 and 18 months, restricting prescriptions to specialists in neurosurgery and child rehabilitation. The results determine when to end the therapy. The usual duration of treatment, considering the different ages at which children are referred, is usually around 4 months, but if the treatment begins between the ages of 5 and 7 months, most infants who need a cranial orthosis usually take 2 months to complete it. This is a period in which the child must wear a helmet 24 hours a day, removing it only for daily cleaning. After one year of age the therapeutic benefit of the orthopaedic correction is already slight. After 15 months, the possibility of orthopaedic correction is practically inexistent.

We must insist on the proven fact that primary prevention and rehabilitation treatment applied well makes it unnecessary to recommend the orthopaedic helmet in most cases.

### **3. Surgery.**

Surgery is always the last resort. As regards plagiocephalies, taken to extreme cases, it is to be restricted to craniostenotic plagiocephalies.

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## Guidelines for monitoring and referral from primary care

(table 4)

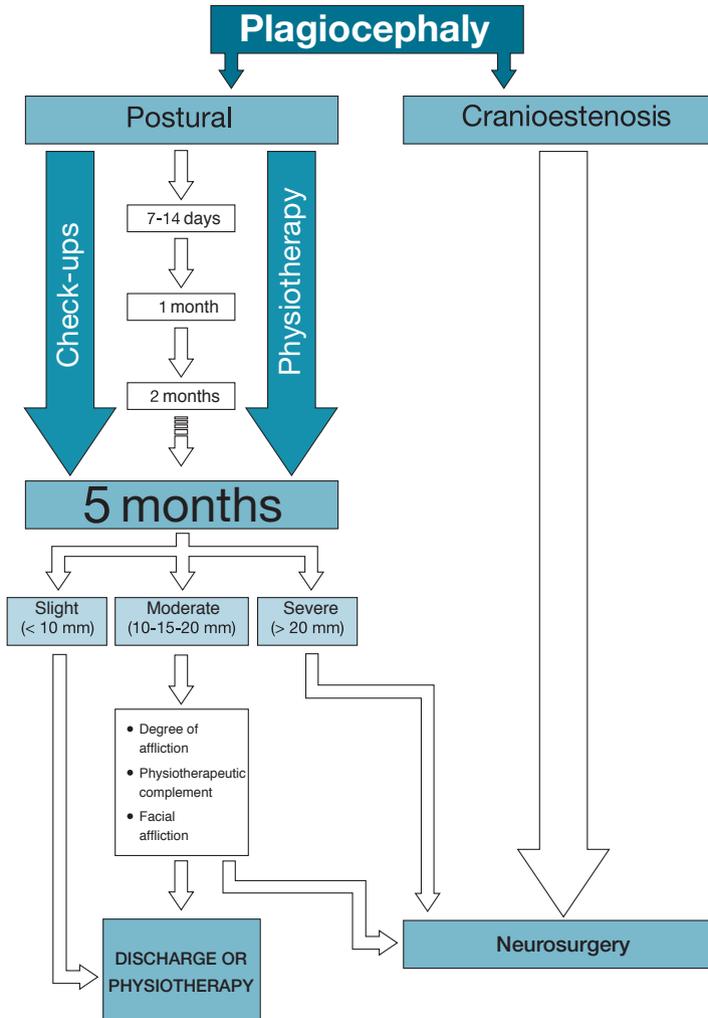


Tabla 4 - Guidelines for monitoring and referral from primary care.

- Any suspected craniostenosis must be referred without delay to neurosurgery, indicating the reasons for the suspicion in order to facilitate the response from the hospital.
  - Once the diagnosis of positional plagiocephaly has been established, the family is to be shown how to apply muscle stretching and positional manoeuvres for torticollis. For this purpose, we have drawn up some prevention and treatment pamphlets based on prior initiatives (41) in order to disseminate the preventive measures and bolster corrective practices. (fig. 27 and fig. 28)
  - A week or fortnight after the visit in which the diagnosis was made, an appointment must be made with the family to check on the rehabilitation measures and strengthen the confidence of parents and carers in their effectiveness and their ability to carry them out.
  - Once it is certain they are being done properly, there is to be a monthly check-up in which cephalometric and plagiocephaly indices are measured, along with the usual determination of the cranial perimeter to check on the deformation's progress.
  - The limit for the monthly check-ups and the time to take the decision on referral to neurology is set at 5 months of age. As of this age, the rehabilitation measures begin to lose their effectiveness due to the infant's autonomy of head movement. Any more delay may have a negative effect on achieving complete correction should orthopaedic treatment be needed.
  - The decision on referral to neurosurgery shall depend on:
    1. Assessing the severity of the deformation: severe forms ( $PI > 20$  mm) must be referred without delay. Moderate forms ( $IP$  10 to 20 mm) should be decided by assessing the degree of affliction, the extent of compliance with the positional measures and rehabilitation exercises. Slight forms ( $IP < 10$  mm) without craniofacial affliction are not generally considered to need orthopaedic treatment; one must insist on the positional measures.
    2. Facial affliction is more difficult to correct and should be specially assessed without adhering strictly to solely anthropometric considerations, although this is more commonly associated with severe forms.
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With the collaboration of:

