

# Orthotic treatment improves cranial base abnormality in patients with craniofacial microsomia and deformational plagiocephaly

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## SUMMARY

**Introduction:** Craniofacial microsomia is associated with hypoplasia of the facial skeleton and musculature. These primary defects cause a secondary alteration of the craniofacial skeleton. Current therapies do not attempt to correct the cranial base deformity in childhood. Another cause of oblique deformities of the skull is deformational plagiocephaly. This common disorder is secondary to external deformational forces and tends to improve with time and may require only conservative treatment. **Method:** We present two cases of deformational plagiocephaly superimposed upon hemifacial microsomia. Orthotic treatment was utilized to improve both the deformational plagiocephaly and the cranial base deformity. **Conclusion:** This novel therapy has the potential to correct the cranial base deformity in craniofacial microsomia.

**Descriptors:** Skull/abnormalities. Facial asymmetry. Orthotic devices. Plagiocephaly.

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## INTRODUCTION

Craniofacial microsomia is seen in 1/5000 live births<sup>1</sup>. It is often associated with a variety of malformations including mandibular hypoplasia and associated changes in facial musculature as well as maxillary hypoplasia<sup>2,3</sup>. An associated feature of this condition, concurrent with hypoplasia of the mandible, is an alteration of the cranial base, which appears to be related to the asymmetric growth of the mandibular ramus<sup>4</sup>. Previous treatments of this condition have centered on attempts to alter the mandibular ramus and maxillary position. Longer-term management has focused on attempts to correct various elements of the secondary deformities (i.e. mandibular, maxillary or soft tissue)<sup>5</sup>. No previous treatments have addressed the cranial base abnormality and asymmetry in infancy. This case report describes the first treatments of the cranial base abnormality in infants with craniofacial microsomia and deformational plagiocephaly.

## METHODS AND RESULTS

The first patient of this serie was a three months old infant boy who presented with left sided craniofacial microsomia. His initial presentation included superimposed

right-sided deformational plagiocephaly. The patient had been back sleeping and the patient developed significant deformational plagiocephaly (Figures 1A and 1B). Given the degree of deformational plagiocephaly it was elected to treat this with dynamic orthotic cranioplasty. A three dimensional CAT scan (Figure 1C) was obtained prior to use of the DOC Band® and pre and post treatment skull molding (molds) were used to ascertain endpoints (Figure 1D). Post treatment results have demonstrated excellent correction of cranial base portions as well as change in the zygomatic prominence laterally from the pre-treatment position (Figures 1E and 1F). Four years follow-up has demonstrated the correction of head shape (Figures 1G and 1H).

The second patient has 4 week old female, was born of a full term vaginal delivery with no complications during the pregnancy, who presented left craniofacial microsomia, including mandibular hypoplasia and microtia. One month later, she was noted to have an abnormal skull configuration, consistent with deformational plagiocephaly and secondary torticollis. Physical examination revealed a deformed skull with a right occipital and left frontal flattening. There was compensatory bulging in the right frontal and left occiput. In planning treatment, we elected to attempt correction of

**Figure 1** - Case 1. A three months old infant boy with left craniofacial microsomia. **A.** Pre operative frontal view. **B.** Vertex view. **C.** Pre-operative 3D CAT scan. **D.** Orthotic treatment. **E.** Immediate post treatment frontal view. **F.** post treatment vertex view. **G** and **H.** Four year follow-up.



the cranial base position simultaneously with the DOC band. She started to use the helmet with 7 months of age, until 13 months. The skull shape after the treatment had improved.

## DISCUSSION

The association between craniofacial microsomia and plagiocephaly is not rare. Padwa et al.<sup>4</sup> presented 15 cases of frontal flattening, including 14 with characteristic deformational abnormality. These represented 10% of their cases. Torticollis was noted in 8 out of 14 cases. They discussed this as possibly coincidental with different etiologies and pathogenesis, or as potentially causally related. It is possible that the muscular hypoplasia in craniofacial microsomia may not permit the appropriate position of the cranial base. Also, the cervical spine may have undetected anomalies that contribute with the deformation.

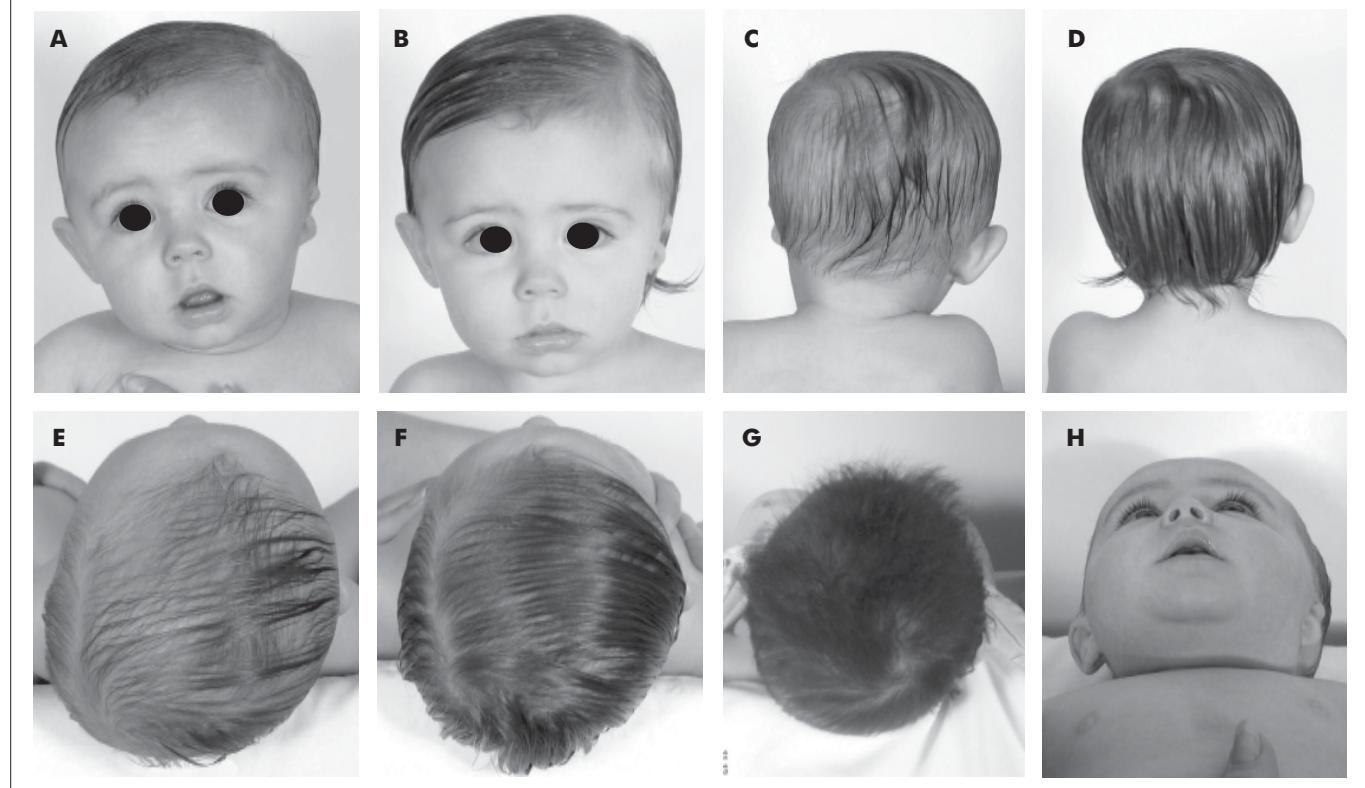
Treatment strategies for patients with craniofacial microsomia have centered on management of the primary maxillary or mandibular hypoplasia, once established in the early childhood or late childhood period. However, few non surgical treatments exist for management of skull deformities in infancy, primarily because no satisfactory means existed to provide such treatment. Since the advent of treatment of severe deformational or positional plagiocephaly with the DOC Band<sup>6</sup>, it has occurred to our

team that we may be able to manage significant alteration in the cranial base morphology by modest means to affect significant changes in condylar portions and affect potentially longer lasting changes in condylar position.

While clearly too early to ascertain significant changes in the cranial base long term, we anticipate that early molding of the cranial base may have a significant and potentially beneficial effect on the condylar position, potentially ameliorating alteration in the divergent condylar position. This may prove beneficial in management of positional anomalies such as the microtic ear. We feel, as well, that deformational plagiocephaly which has been increasing in frequency as the "Back to Sleep" campaign has progressed<sup>7-10</sup>, may continue to provide diagnostic challenges in mild cases of hemifacial microsomia. But of greater concern is that such cases of deformational plagiocephaly superimposed upon hemifacial microsomia now present an even greater therapeutic challenge. Further alteration of the cranial base position as exacerbated by deformational plagiocephaly (especially if it is on the ipsilateral occipital region) potentially may severely aggravate this condition.

For this reason we feel that aggressive early intervention of deformational plagiocephaly in hemifacial microsomia is warranted. Such alteration in cranial base abnormalities may yield significant alteration, which cannot be appropriately addressed without proper orthotic intervention.

**Figure 2** - Case 2. Four week old female with left craniofacial microsomia. **A.C.E.** and **G**. Pre operative views. **B.D.F.** and **H**. Post treatment view, 6 months after started using the helmet.



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