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Methods paper

Hereditary premature closure of a coronal suture in the Abraham Lincoln family

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ABSTRACT

The most easily recognized facial features of unilateral premature closure of a coronal suture in the skull are an upward arching of the superior orbital rim and a smaller face on the involved side. Photographs indicate that at least 9 individuals over 5 generations of the Abraham Lincoln family showed this anomaly.

1. Introduction

Normal development of the calvarium involves five major suture systems, three of them paired (coronal, lambdoid, and squamosal) and two single (sagittal and metopic). Normal growth of bone occurs perpendicular to the course of the suture, ceasing with closure of the suture at various times in adolescence or even later. Premature closure, or craniosynostosis, may occur as early as prenatal or perinatal life, producing marked abnormality, or late in development, producing milder problems. Craniosynostosis appears in approximately 1 in 2500 infants. Most cases are sporadic, but of those with a genetic component, there appears to be a predilection for involvement of the coronal suture. The coronal suture, both right and left parts, divides the anterior cranial segment from the middle cranial segment and is overall the second most common suture involved, occurring in 20% to 30% of craniosynostosis patients. In those cases where primarily only one suture is involved, the resulting deformity is more predictable than in cases involving two or more sutures (Cohen, 1980; Cohen, 2009; Cohen and MacLean, 2000). Involvement of a single coronal suture is 4–7 times as frequent as bilaterality, the right side twice as often affected as the left, and females twice as often as males (Di Rocco et al., 2012).

Premature closure of 1 coronal suture, also termed synostotic frontal plagiocephaly, is associated with the following constellation of facial anomalies, among others:

1. A relative elevation or arching of the superior orbital rim or brow ridge due to failure of full development of the frontal bone (Fig. 1). In severe cases with early closure, this feature may be accentuated by a compensatory overdevelopment of the contralateral forehead with depression of its upper orbital rim. The sphenoid wing on the synostotic side, as it forms the orbital roof, is also elevated and, on a radiographic frontal projection of the skull, produces an upswept diamond-shaped oblique configuration suggestive of the traditional diamond pattern of the theatrical harlequin costume. This is the "harlequin eye" deformity described by radiologists (Fig. 2). (http://www.medcyclopaedia.com/library/topics/volume_vii/h/harlequin_eye/gharlequin_eye_fig1.aspx?s=harlequin+eye&mode=1&syn=&scope=)
2. Decrease in size of the ipsilateral face producing facial asymmetry. Although facial asymmetry *per se* is so common as to be considered normal, when it is marked or especially when it is associated with arching of the ipsilateral brow it may be considered of diagnostic significance
3. A smaller orbit on the involved side with rounding of the palpebral fissure because of upper displacement of the superior lid sulcus, often with narrowing of the horizontal diameter
4. Elevation and anterior displacement of the ipsilateral ear
5. Comparative flattening of the ipsilateral forehead, sometimes with prominence (or "bossing") of the contralateral forehead, producing an oblique deformity of the skull (frontal plagiocephaly)

2. Methods

Photographs of Lincoln and his family were examined, as well as two life masks of Lincoln made in 1860 and 1865. Horizontal linear measurement of the two half-faces was done to reveal facial asymmetry, as well as placement of horizontal parallel lines across the

Abbreviation: DNA, deoxyribonucleic acid.

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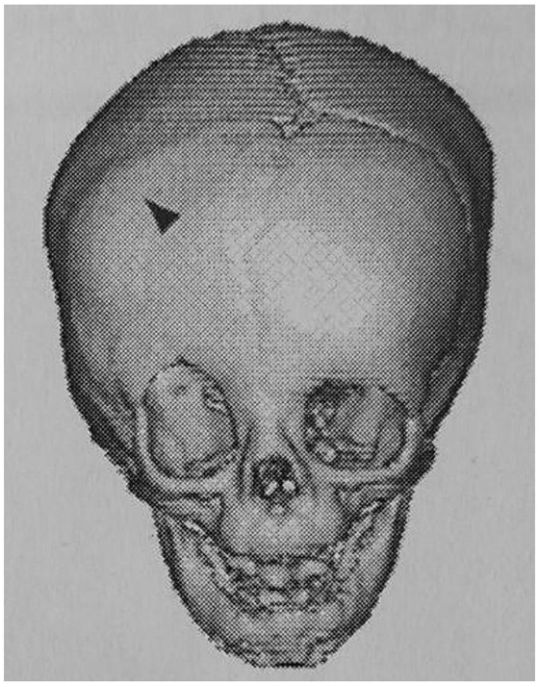

Fig. 1. Infant skull with a synostosis of the right coronal suture, showing the smaller face on that side together with elevation and arching of the superior orbital rim (from multiple CT reconstructions by M. Vannier as shown in Cohen and MacLean [p123], by permission of the authors).

face to reveal vertical displacement of orbits and ears on the two sides. In addition, three dimensional laser scanning of the masks was done in order to document the facial configuration (Fishman and DaSilviera, 2007).

3. Results

Table 1 shows the names and relationships of 12 members of the Lincoln family over 5 generations.

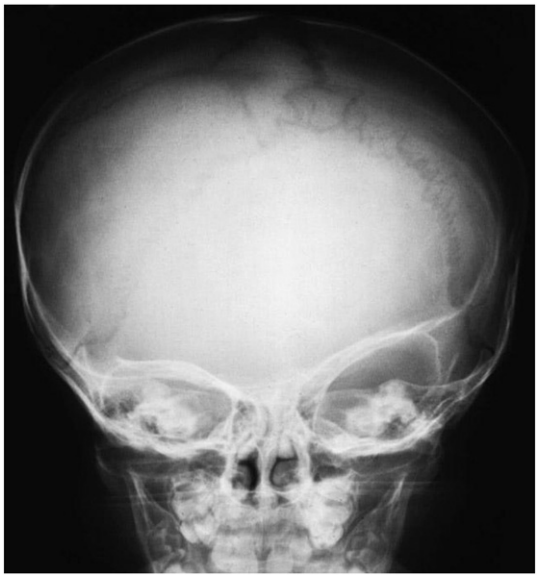

Fig. 2. Right harlequin eye deformity: skull radiograph showing elevation of the right sphenoid wing and orbital enlargement due to a premature fusion of the right coronal suture (available at: http://www.medcyclopaedia.com/library/topics/volume_vii/h/harlequin_eye/gharlequin_eye_fig1.aspx?s=harlequin+eye&mode=1&syn=&scope=Accessed October 5, 2009).

Table 1
Lincoln family tree.

Generation I
Thomas Lincoln (1758–1851) — (L)
Generation II
Abraham Lincoln (1809–1865) — (L)
Generation III
Robert (1843–1926) — (L)
Edward (1846–1850) — (L)
William (1850–1862) — (L)
Thomas (1853–1871) — (L)
Generation IV
Mary (1869–1938) — (R)
Abraham II (1873–1896) — (L)
Jessie (1875–1948)) — (?)
Generation V
Lincoln Isham (1892–1971) — (?)
Mary Beckwith (1898–1975) — (?)
Robert Beckwith (1904–1975) — (L)

The side in which synostosis became manifest is indicated in parentheses after each name. A question mark denotes the subject where no photograph was found adequate to determine whether synostosis was present.

3.1. Abraham Lincoln (1809–1865)

Photographs of the face may show bilateral asymmetry when the angle of view is in a true frontal plane. Shifting the angle of view even slightly to one side of the median quickly masks the asymmetry. Several true frontal photographs of Abraham Lincoln exist, the most often reproduced being the one taken in November 1863 (Fig. 3). Facial asymmetry is marked, with a smaller left orbit and a left brow appearing to be elevated compared to its fellow. This arching of the left brow gives the face a somewhat quizzical or skeptical look, as if he were using the frontalis muscle to raise one eyebrow. Its appearance in the life masks (where the bony contours are not masked by the eyebrows) shows it not caused by muscle action but rather by a difference in the configuration of the bone of the orbital rims (Fig. 4). This mask also shows elevation and forward placement of the left ear. All these signs are consistent with a left coronal craniosynostosis.

Lincoln experienced occasional diplopia and was noted at times by others to show an upward deviation of the left eye, and this is also manifest in the 1863 photograph. Several photographs also show him tilting his head to the right (Goldstein, 1997). This was most likely due to an underaction of the left superior oblique muscle due to mechanical factors

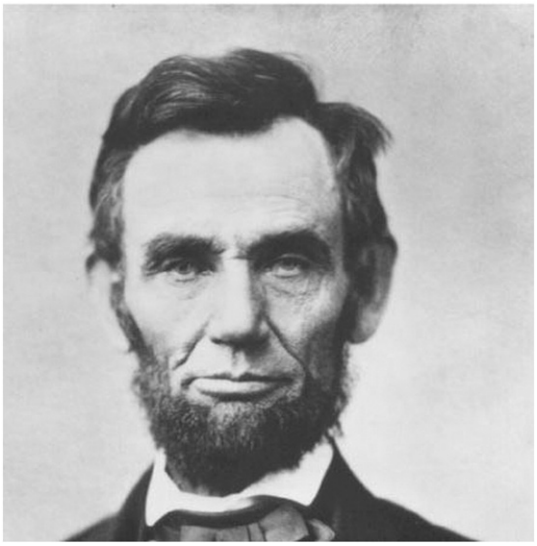

Fig. 3. Frontal photograph of Lincoln from 1863 (photograph courtesy of Abraham Lincoln Museum of Lincoln Memorial University, Harrogate, TN).