

Mercedes Benz craniosynostosis

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A 2-year-old girl presented with an abnormal shaped head since early infancy. Her growth and development were normal. A computerized tomography (CT) scan showed a fusion of sagittal and bilateral lambdoid sutures, typical of Mercedes Benz pattern craniosynostosis [Figure 1]. Except for widened subarachnoid spaces in the frontal region, she did not have any brain anomalies.

Mercedes Benz pattern craniosynostosis is also known as bilateral lambdoid and sagittal synostosis (BLSS). It was first described in 1976 by Neuhauser in a series of seven patients with synostosis of the sagittal and lambdoid sutures, short stature, and developmental delay.^[1] The term “Mercedes Benz” syndrome was coined by Moore in 1998 due to the characteristic appearance of the fused sutures on three dimensional CT imaging.^[1]

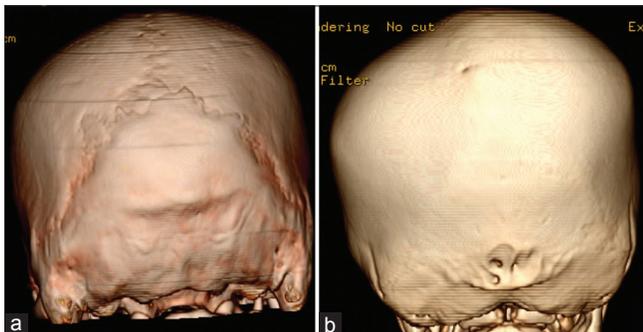


Figure 1: (a) Computerized tomography scan with surface shaded display of a child with normal brain imaging showing normal sagittal and both lambdoid sutures, (b) Computerized tomography scan of patient showing fusion of sagittal and both lambdoid sutures typical of Mercedes Benz pattern

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BLSS is a rare disorder characterized by invagination of the occipital squama resulting in a step-like deformity of the occiput, and a typical head shape described as anterior turricephaly with mild brachycephaly.^[2] Locally marked growth restriction is evident in the posterior fossa with the compensatory secondary expansion of the anterior fossa manifesting a degree of frontal bossing which mimics bicoronal synostosis.^[3] The incidence of BLSS is 1.4%.^[4] BLSS is a heterogeneous disorder with syndromic, chromosomal, and isolated forms.^[1] The associated anomalies include the Chiari I malformation (CM), hydrocephalus, and venous anomalies.^[2] Although, the appearance of frontal bossing could lead to inadvertent surgery in the frontal region, attention to the occipital region with wide early suture excision and vault shaping is indicated.^[3] Children, in addition, require foramen magnum decompression if CM is present.^[2]

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Conflicts of interest

There are no conflicts of interest.

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