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# 벤츠 패턴을 가진 두개골 조기 유합증: 증례보고

류석태 · 임소영 · 문구현

성균관대학교 의과대학 삼성서울병원 성형외과학교실

## Mercedes Benz Pattern Craniosynostosis: A Case Report

Suktae Ryoo, M.D., So Young Lim, M.D.,
Goo Hyun Mun, M.D.

Department of Plastic Surgery, Samsung Medical Center, Sungkyunkwan University, School of Medicine, Seoul, Korea

**Purpose:** Craniosynostosis of three or more cranial sutures was not common. "Mercedes Benz pattern," named by Moore1 was a rare form of craniosynostosis and had an atypical pattern of premature closure of cranial suture. It was not reported in Republic of Korea. We report this case with literature review.

**Methods:** A 13-months-old male patient visited our clinic due to exophthalmos. He showed normal developmental course. Other neurological tests were normal but he was Crouzon syndrome patient. CT scans showed bilateral lambdoid and posterior sagittal sutures were fused and the length of the skull was extended. Cranioplasty with pi craniotomy & Barrel-Stave osteotomy and recombination of the bone flap was performed.

**Results:** The patient was discharged after post operative 10 days without any complications. In follow up visit after 2.7 years, he was in good state without recurrence and functional abnormality of skull.

**Conclusion:** This was the first case of Mercedes Benz pattern craniosynostosis with Crouzon syndrome in Korea. This type of craniosynostosis has to be considered differently from single type of craniosynostosis or typical syndromic craniosynostosis clinically and surgically.

**Key Words:** Mercedes Benz pattern, Multiple craniosynostosis, Syndromic craniosynostosis

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**Address Correspondence :** So Young Lim, M.D., Department of Plastic Surgery, Samsung Medical Center, Sungkyunkwan University School of Medicine, 50 Iiwon-dong, Gangnam-gu, Seoul 135-710, Korea. Tel: 02) 3410-2235/Fax: 02) 3410-0036/ E-mail: sy72.lim@samsung.com

## I. INTRODUCTION

Craniosynostosis is the congenital disease that one or more sutures of calvarium and cranial base were fused early before the normal time so growth inhibition of the brain, increased intracranial pressure and craniofacial deformities can be found. The single isolated sutural fusion is rarely associated with syndromes. The majority of multiple sutural craniosynostosis have identifiable syndromes which affect mainly both coronal sutures. Multiple sutural fusions not involving bilateral coronal sutures are rare. Syndromic craniosynostosis which involve not coronal but sagittal and lambdoid sutures was not reported yet. "Mercedes Benz pattern," named by Moore1 which means the premature fusion of bilateral lambdoid and sagittal sutures has not been reported in Republic of Korea. So we report this case with literature review.

## II. CASE

A 13 month old male patient visited our clinic due to exophthalmos (Figs. 1, 2). He was born in gestational age 39 + 3 weeks with normal full term spontaneous delivery. Bilateral optic disc elevation and edema were detected by the ophthalmologist. MRI and CT scan images showed frontal bone bossing, prolonged AP diameter, clivus flattening, increased digital marking, maxillary hypoplsia and presumed Chiari malformation type I with cerebellar tonsilar herniation. Also intracranial pressure was increased. His mother reported the patient suffered from rough breathing, especially during sleep accompanied by shortness of breathing symptoms and could not sleep well. He was in normal developmental course in activity and speech. Other neurological tests were normal. His father's appearance showed some features of Crouzon syndrome such as exophthalmos and midface hypoplasia. On admission, his weight was 11.1 kg (75 percentile), the height was 76 cm (25 pecentile), head circumference was 46 cm (50 percentile),

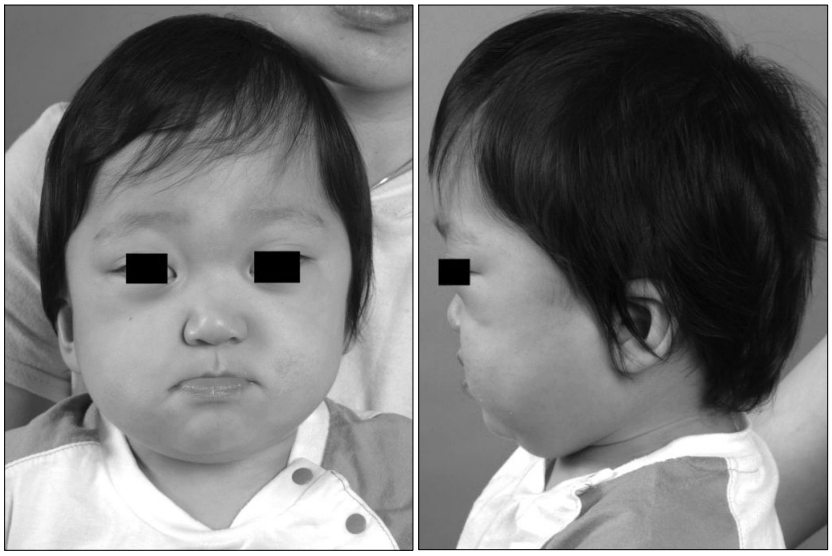

**Fig. 1, 2.** Pre operative medical photographs.

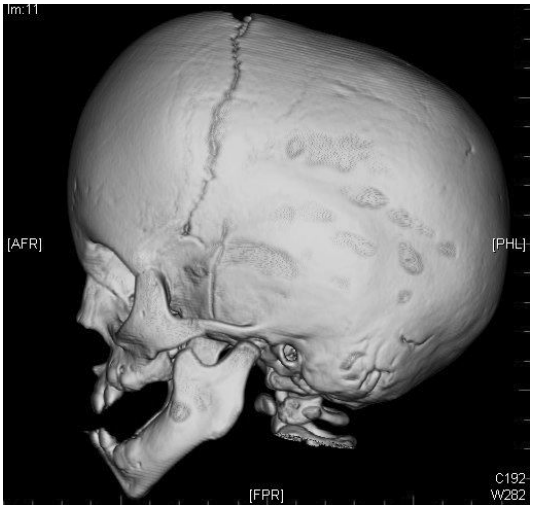


**Fig. 3.** Lateral CT image. This picture showed premature fusion of lambdoid suture and frontal bulging due to compensatory expansion and maxillary hypoplasia.

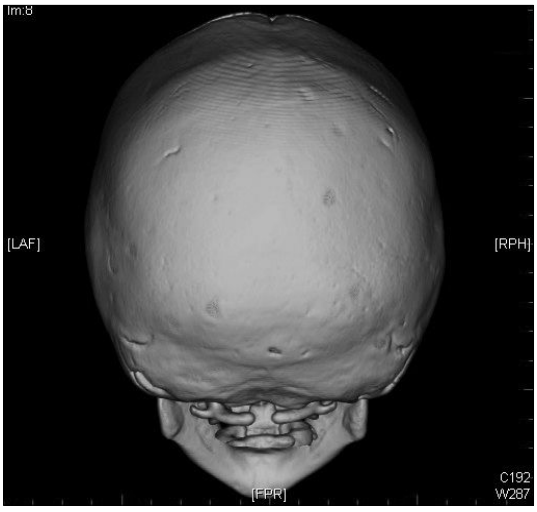


**Fig. 4.** Posterior CT image. We can find Mercedes Benz pattern's premature closure of bilateral lambdoid and posterior sagittal sutures.

diameter of anterior to posterior was 16 cm and BPD (biparietal diameter) was 12.6 cm, respectively. CT scans showed bilateral lambdoid and posterior sagittal sutures were fused, the length of the skull (AP diameter) was extended, inner calvarial digital marking was increased and clivus flattening and superior bulging of the frontal region were found. Also maxillary hypoplasia was detected (Figs. 3, 4). These findings suggested "Crouzon syndrome". Cranioplasty was performed. The bone flap was obtained through pi craniotomy & Barrel-Stave osteotomy and recombination of the bone flap was performed (Fig. 5). The patient was observed in ICU for 3 days and discharged after 10th post operative days. By follow up visit postoperative 2.7 years, his head

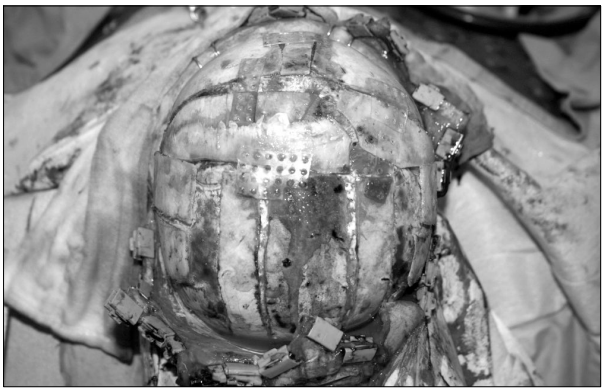

**Fig. 5.** The bone flap was obtained through pi craniotomy & Barrel-Stave osteotomy Recombination of the bone flap was performed.

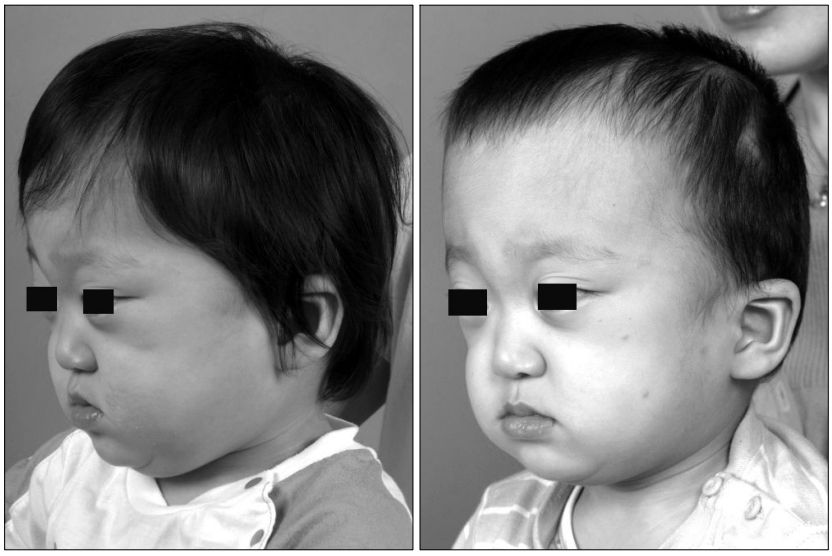

**Fig. 6.** Pre operative and Post operative 83 days oblique medical Photographs. Frontal bulging and upslanting was improved.

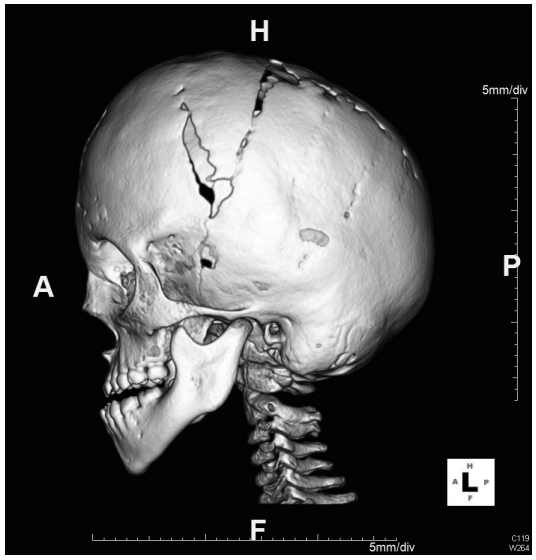


**Fig. 7.** Postoperative 2.7 year-follow up lateral CT image. We could find improvement of frontal bossing and reduced relative AP diameter. The vestige of pi-craniectomy and remnant fused sutures were also found.

circumference was 50 cm, AP diameter was 16.5 cm and biparietal diameter was 14.3 cm. CT findings showed improvement of frontal bossing and he was well state without any recurrence and functional abnormality of skull (Figs. 6, 7).

## III. DISCUSSION

According to Virchow's law, skull is expanded in parallel to the suture of premature closure by the principles of hyperexpansion and compensation. Accordingly, premature closure of metopic suture can form trigonocephaly that of bilateral coronal sutures cause brachycephaly, in case of premature closure of sagittal suture, there can be scaphocephaly. Also premature closure of unilateral coronal or lambdoid suture is a form of plagiocephaly. The prevalence of craniosynostosis was estimated to be one per 1,800 to 2,200 births.[7] Most of non-syndromic craniosynostosis is due to isolated single suture fusion. The single isolated suture fusions are rarely associated with syndromes.[1] Premature fusion of two sutures were usually related to typical syndrome. In cases of more than two sutures, according to the Chumas in 1997, 17 patients were reported who had three or more skull sutures fused in 1474 patients of craniosynostosis. Of these, 10 patients had premature closure of bilateral lambdoid and sagittal sutures and were classified nonsyndromic complex type of craniosynostosis.[3,4] Moore named this pattern to "Mercedes Benz pattern" with 3 patients in 1998.[1] In 2007, Rodney reported 2 case of Z-pattern's premature closure (left coronal, sagittal, right lambdoid)and they showed nonsyndromic pattern.[6] Rhodes reported 11 cases of Mercedes Benz pattern craniosynostosis in 2010.[2] Usually, fusion of two sutures involves the right and left coronal sutures.[1] The majority of these bilateral coronal fusions occur as a component of an identifiable syndrome such as Apert, Crouzon, Pfeiffer, and so on. Rhodney reported that Z-pattern craniosynostosis did not appear to be associated with any known genetic markers and he did not identify any other phenotypic anomalies outside of the skull and brain.[6] And Rhodes reported two cases showed an association with atypical syndrome,

one was Opitz syndrome the other Potocki-Shaffer syndrome. He reported there was no intelligence problem except Opitz syndrome patient. 2/3 patients of Benz pattern showed cerebellar tonsillar herniation and more than half of them were reported on the need for surgical decompression.[2] Except these two patient, others showed nonsyndromic pattern. The patient who showed Opitz syndrome had premature fusion of unilateral coronal suture additionally. Thus, Most of Mercedes Benz pattern's craniosynostosis showed nonsyndromic pattern. But in our case, the patient was suggested as Crouzon syndrome. The patient showed normal developmental growth except the problems caused by skull deformity. Also intelligent problem was not to be seen. Findings on MRI showed Chiari malformation type I and apnea findings were also shown as stated in previous reports. These findings suggested Crouzon syndrome. As we reviewed, we thought that this rare form of complex craniosynostosis presented a clinical entity distinct from the single isolated craniosynostosis or typical syndromic craniosynostosis which involved two sutures. This multiple craniosynostosis results in deformity of other skull area and also can cause life threatening condition such as cerebellar tonsillar herniation.[8] This combination of multiple sutures' craniosynostosis envelops the posterior cranial fossa with three dimensional growth restriction in this area so cerebellar tonsillar herniation can happen more frequently than in other type of craniosyniostosis. Also compensatory growth and expansion of the brain in the anterior fossa resulted in marked frontal bulging and upslanting, and drawing upwards of the orbital roofs. So correction for this pattern of craniosynostosis needs to be quite different from that of an isolated sagittal synostosis, with skull elongation instead of shortening.[1,2,5]

As shown, we present the first patient with Mercedes Benz pattern craniosynostosis associated with Crouzon syndrome in Korea. The collection of more cases and research about this type of craniosynostosis will be needed.

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