Case Report

CLOVERLEAF SKULL DEFORMITY

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ABSTRACT

Cloverleaf skull is a rarely seen deformity and the prognosis is poor in untreated cases. The main components of the deformity are hydrocephalus, trilobed head shape, shallow orbits, and midfacial hypoplasia. It may be seen either as an entity or in association with other syndromes, such as Apert's, Crouzon's, Pfeifer's syndromes, and skeletal dysplasias. The presented case is in association with some skeletal anomalies, together with ambiguous genitalia and dermal sinus tract over lumbar spine. As early surgical intervention is suggested to prevent the damages of increased intracranial pressure, we operated the patient on the second day of her life. Although the vital signs of the infant returned to normal limits immediately after the operation, the patient died on the third postoperative day because of diffuse intravascular coagulopathy.

Key Words: Cloverleaf deformity, Kleeblattschadel, Craniostenosis, Congenital dislocation of the hip.

INTRODUCTION

The name "Kleeblattschadel" or "cloverleaf skull" for the grotesque configuration of trilobar skull was first given by Holtermüller and Wiedemann (1). Major craniofacial manifestations of cloverleaf skull deformity (CSD) were listed as follows: 1. Cloverleaf skull, 2. Severe exophthalmos, corneal ulcerations, hypertelorism and shallow orbits. 3. Downward displacement of the ears, occasionally reaching a position parallel to the shoulders. 4. Beaked nose with a depressed nasal bridge. 5. Premaxillary and maxillary hypoplasia with relative prognatism and occasionally macrostomia, macroglossia etc. (2,3). Although this deformity can be seen as a single pathology, it may also be seen as an element of a syndrome, or in combination with other skeletal anomalies. CSD is etiologically and genetically heterogeneous and may occur in association with a number of disorders which result from genetic mutations such as hypochondroplasia (4). Prenatal diagnosis of the skull abnormality and accompanying skeletal abnormalities is possible using ultrasonography (5,6).

CASE REPORT

This female neonate was born to a 26-year-old gravida I, para 1 woman at 39 weeks of gestation with routine vaginal delivery. The course of pregnancy was uneventful, but an ultrasonographic evaluation two weeks before the delivery revealed an abnormal shape of cranium without any specific diagnosis. The APGAR score of the infant was 10 at birth. The same day she was referred to the Pediatrics and Neurosurgery Departments, for investigation of her apparent craniofacial deformity.

Her weight was 3000 g and head circumference was 39 cm. The trilobed appearance of her skull with bulging of frontal and both temporal regions was diagnosed as "Cloverleaf skull deformity". There was hypertelorism, midfacial hypoplasia and bilateral exophthalmos. Both ears were low-placed and the nasal root was flattened (Figs 1a, b).

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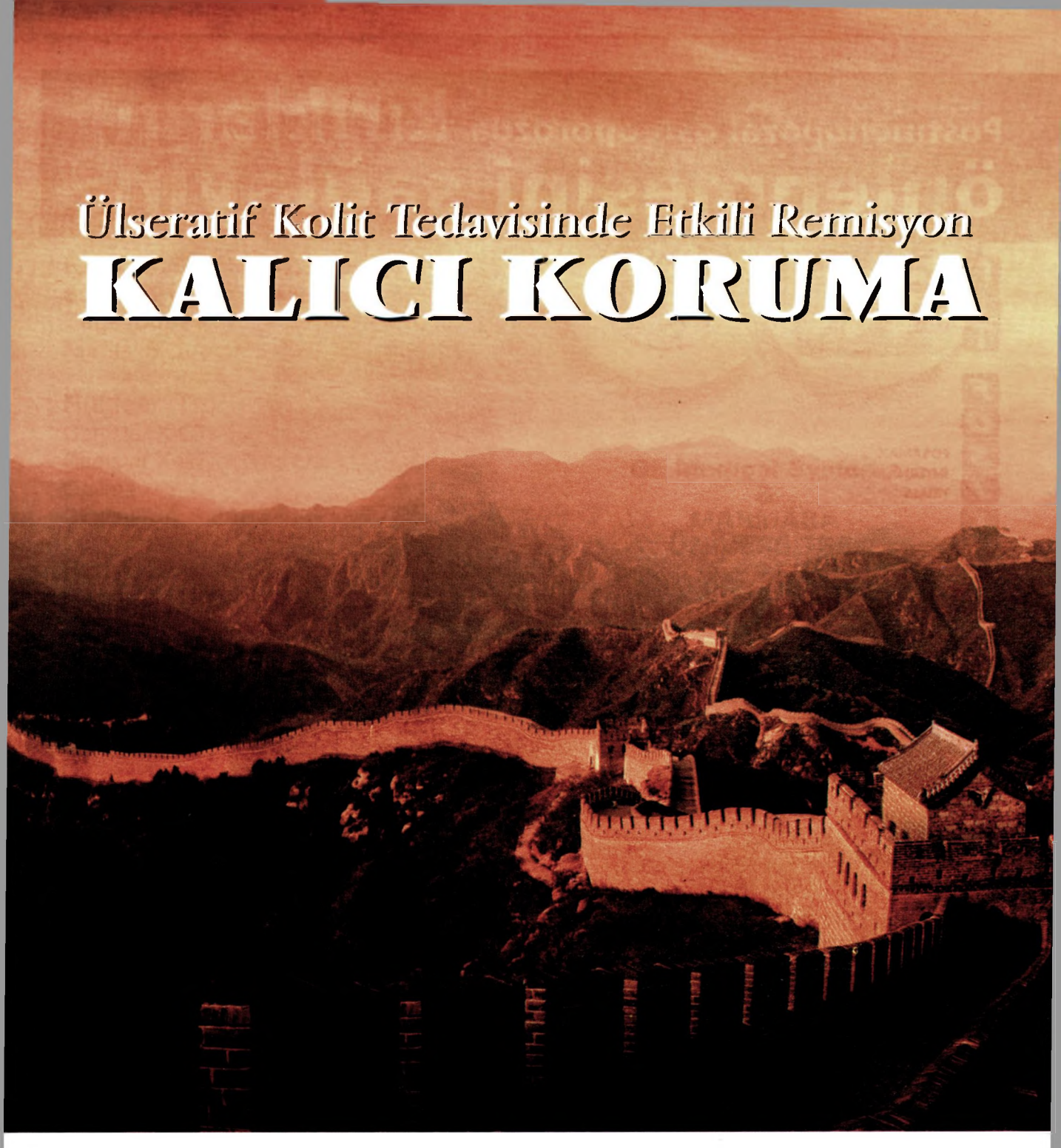

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1. *Black DM et al for the Fracture Intervention Trial Research Group. Randomised trial of effect of alendronate on risk of fracture in women with existing vertebral fractures. Lancet 1996; 348: 1535-1541*
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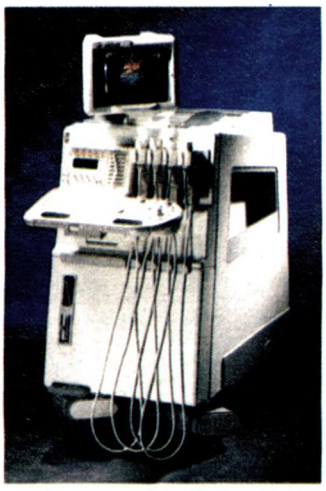

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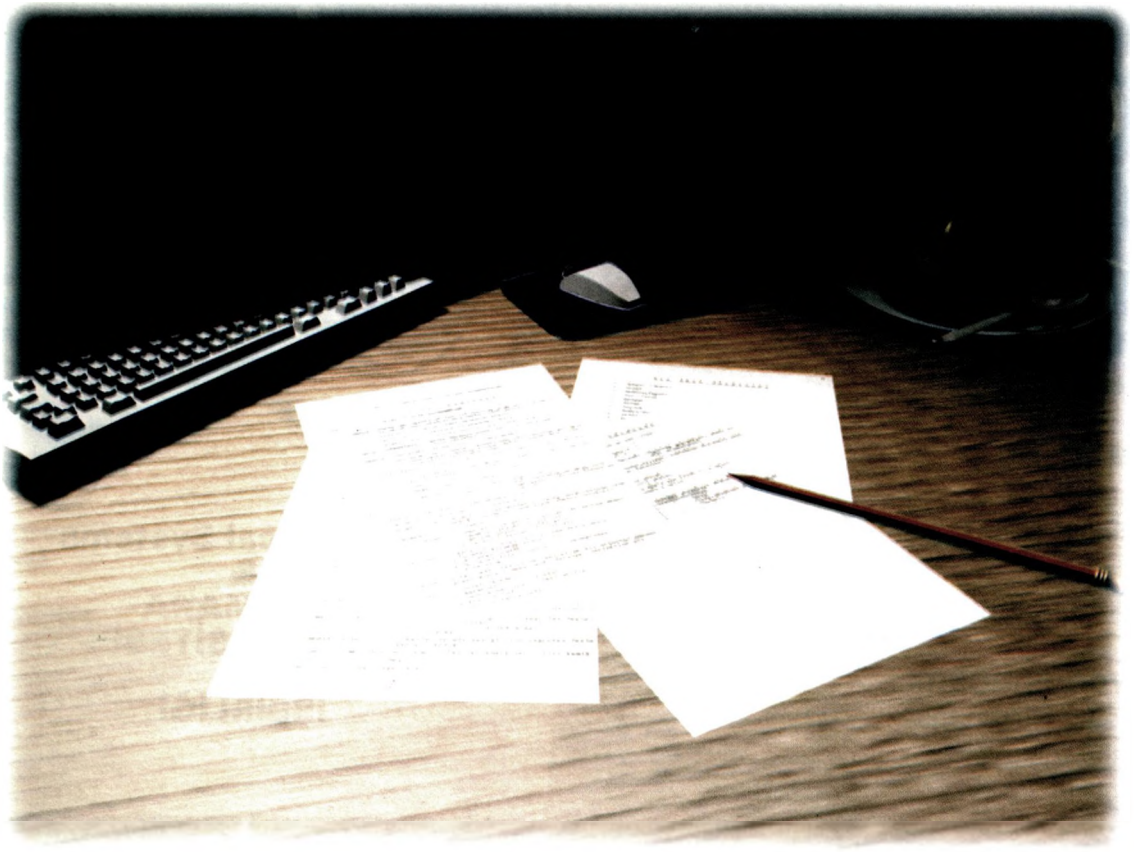

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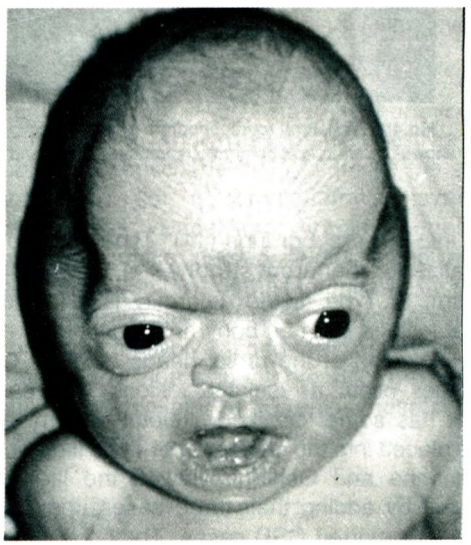
She had abnormal external genital organs, diagnosed as ambiguous genitalia. There was a dermal sinus tract over the lumbar area in the midline. The shape and length of her extremities and fingers were normal.

Cranial plain roentgenograms revealed trilobed skull deformity with evident sagittal suture synostosis and honeycomb appearance of calvarial areas (Figs 2 a,b). Cranial computerized tomography scans showed obliteration in all sulci and subarachnoidal cisterns. Ventricular systems were narrowed and posterior lateral ventricles were obliterated bilaterally. Calvarial bones were very thin and irregular. Metopic, squamosal and medial coronal sutures were widened. Orbital cavities were bilaterally shallow. The middle fossa was wide but anterior and posterior fossae were narrow (Fig. 3). The dimensions of the foramen magnum were within normal ranges.

Plain roentgenograms of the whole body were taken. The shapes of the long bones in the extremities were normal. The sacral vertebrae and coccyx showed a lordotic alignment and their tip was palpable (Fig.4).

The day after referral, the patient became tachycardic and tachypneic, because of increased intracranial pressure. The following day, she was operated. After sagittal and coronal synostectomies, frontal and parietal bone flaps were placed on dura with single stitches in the manner of free floating flaps. The occipital bone was so tightly adhered to the dura along the lambdoid sutures that, it was left in place.



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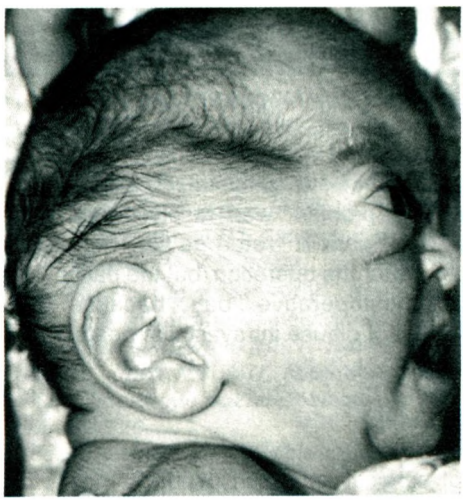

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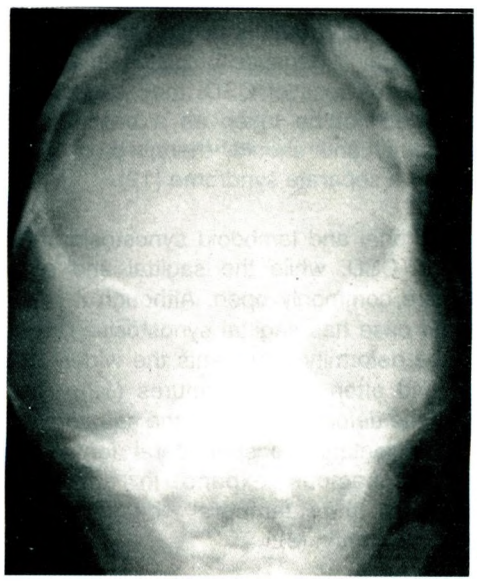
Fig.1: Anterior (a) and lateral (b) photographs of head show the trilobed appearance of head.



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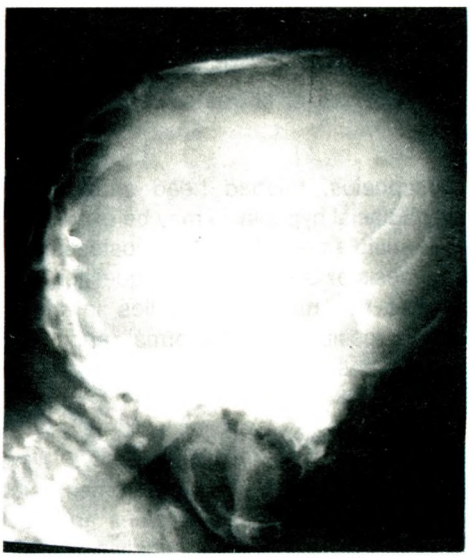

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Fig.2: Anterior (a) and lateral (b) plain cranial roentgenograms. Note the beaten copper appearance of calvarial bones and sagittal suture synostosis.

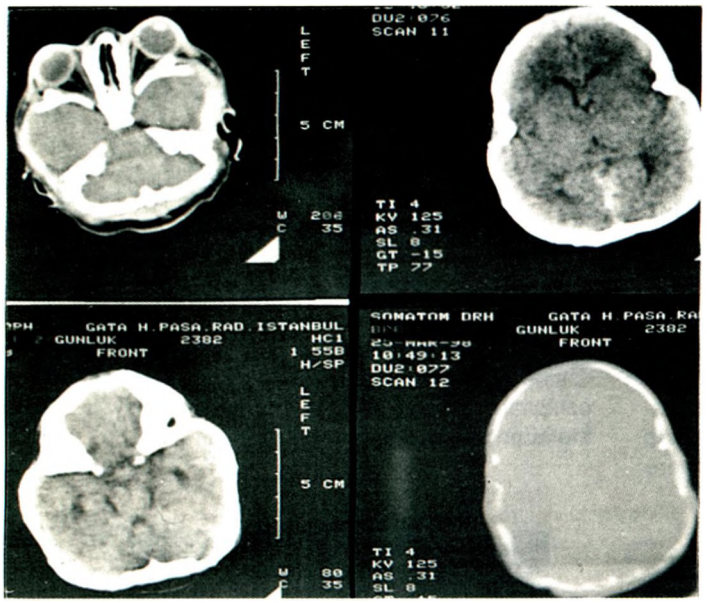

Fig.3: Cranial computerized tomography scans.

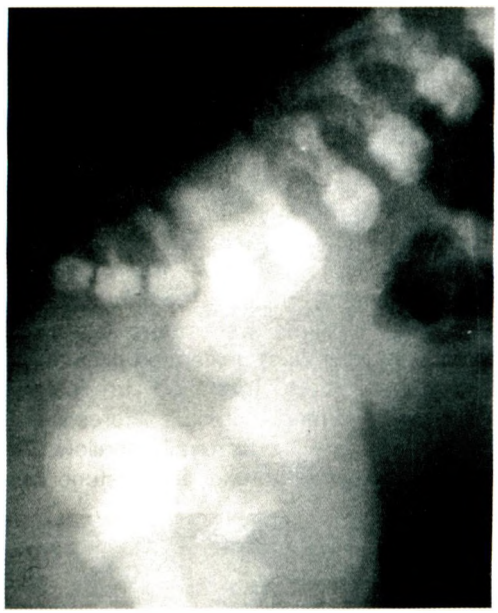

Fig.4: Plain lateral roentgenogram of sacrococcygeal area shows the lordotic alignment of sacral vertebrae and coccyx.

The breathing problem was resolved and the vital signs of the patient returned to normal limits immediately after the operation, but the patient died on the third postoperative day because of the developement of diffuse intravascular coagulopathy.

DISCUSSION

Timing of surgical approach depends mostly on the neurological status of the patients. Although the data concerning the results of early operative intervention of CSD are not enough and it is uncertain whether aggressive decompressive / reconstructive procedures are effective in treating the effects of the malformation on both neurological development and cosmetic appearance, Resnick et al treated their 7 patients in early infancy (7).

The hydrocephalus, trilobed head shape, shallow orbits, and midfacial hypoplasia may be explained with intrauterine multiple cranial suture synostoses, but the real pathogenesis of CSD remains in question (8). It is proposed that the major defect lies in abnormal membraneous ossification. Abnormal endochondral ossification has been suggested as the basis of cranial deformity also (2,9).

CSD rarely occurs as an isolated entity. It mostly occurs in association with other syndromes, such as Apert's, Crouzon's, Pfeifer's syndromes and skeletal dysplasia (10). Partington et al. subdivided the CSD cases into three subgroups (11). Type I is seen with generalized chondrodystrophy of thanatophoric dwarfism. In Type II, the deformity is with localized skeletal lesions such as bony ankylosis of the elbows and subluxation of the radial heads or hips. Type III deformities occur as isolated lesions. The case we present has some localized skeletal deformities and can be placed into the subgroup of Type II deformities. Iannacconne and Gerlini reviewed the literature in 1974 and by adding their three cases, they found a total of 41 reported CSD cases. They concluded that the trilobed deformity is a result of severe alterations in skull development associated with marked hydrocephalus and cranial bulging toward the vertex and bitemporally. Because of quite variable shapes, sizes and evolutions of CSD, they suggest that this pathology cannot be taken as a basis to define a specific entity and should, therefore, no longer be considered a separate syndrome (12).

Mostly, coronal and lambdoid synostosis are usually present in CSD, while the sagittal and squamosal sutures are commonly open. Although it is rare, the presented case has sagittal synostosis. The superior leaf of the deformity represents the widely diastased sagittal and often metopic sutures (12). The lateral leaves of the deformity arise as the squamosal suture, squamosal fontanel, posterolateral fontanel, and/or temporal squamous expand in an attempt to accomodate the temporal dilatation of the hydrocephalic brain (8).

Hydrocephalus is invariably present at birth but develops subsequently in relation to the severity of the premature synostosis (8,13). Whether due to impairment of venous outflow and raised venous pressure in the sagittal sinus or to distruption of cerebrospinal fluid circulation within cortical sulci, both mechanisms specifically acting at the level of the constructive cranial ring, in theory it is possible to avoid the development of hydrocephalus by treating the craniosynostosis early (8). Angle et al. considered the hydrocephalus to be caused by hypoplasia of the chondrocranium with a basilar deformity causing obstruction of the fourth ventricle (2). Prior to the definitive surgery shunting the accompanying hydrocephalus is recommended (14-16). The early computerized tomography scans in our patient did not reveal hydrocephalus and the foramen magnum was seen in normal dimensions. But it was most likely that the communicating hydrocephalus would develop, if the operation were delayed.

The calvarium in CSD has a honeycombed appearance, also known as craniofenestrae which resembles "lückenschadel" (9,17). Unlike the surgical management of conventional syndromal craniosinostosis, paper-thin cranial bones and craniolacunae make surgery tedious and risky in cloverleaf skull because of the high possibility of dural tears. The management of CSD remains a challenge and continues to evolve. Long-term prognosis is still poor, but early surgical intervention provides the potential for relief of craniosynostosis and hydrocephalus (8).

REFERENCES

1. Holtermüller K, Wiedemann HR. Kleeblattschadel-Syndrom. Med Monatsschr 1960;14:439-446.
2. Angle CR, McIntire M, Moore RC. Cloverleaf skull:Kleeblattschadel - deformity syndrome. Am J Dis Child 1967; 114: 198-202.
3. Feingold M, O'Connor JF, Berkman M. Kleeblattschadel syndrome. Am J Dis Child 1969; 118: 589-594.
4. Angle B, Hersh JH, Christensen KM. Molecularly proven hypochondroplasia with cloverleaf skull deformity: a novel association. Clin Genet 1998; 54: 417-420.
5. Boos R, Schmidt W. Prenatal diagnosis of thanatophoric dwarfism with cloverleaf skull. Z Geburtshilfe Perinatol 1986; 190: 225-228.
6. Salvo AF. In utero diagnosis of Kleeblattschadel (cloverleaf skull). Prenatal Diagn 1981; 1: 141-145.
7. Resnick DK, Pollack IF, Albright AL. Surgical management of the cloverleaf skull deformity. Pediatr Neurosurg 1995; 22: 29-37.
8. Lodge ML, Moore MH, Hanieh A, Trott JA, David DJ. The cloverleaf skull anomaly: Managing extreme cranio-orbitofaciostenosis. Plast Recons Surg 1993; 91: 1-14.
9. Burkhardt L, Fisher H. Pathologische Anatomie des Schädels. In: Henke F, ed. Handbuch der Speziellen Pathologischen Anatomie und Histologie, Berlin, Springer-Verlag: 1970: 116-117.
10. Kozlowski K, Warren PS, Fisher CC. Cloverleaf skull with generalised bone dysplasia. Pediatr Radiol 1985; 15: 412-414.
11. Partington MW, Gonzales-Crussi F, Khakee SG, Wollin DG. Cloverleaf skull and thanatophoric dwarfism. Arch Dis Child 1971; 46: 656-664.
12. Iannaccone G, Gerlini G. The so-called "Cloverleaf skull syndrome". Pediatr Radiol 1974; 2: 175-184.
13. Muller PJ, Hoffman HJ. Cloverleaf skull syndrome. J Neurosurg 1975; 43: 86-91.
14. Rogathi M. Cloverleaf skull- a severe form. of Crouzon's syndrome: a new concept in aetiology. Acta Neurochir (Wien) 1991; 108: 45-52.
15. Goh KY, Ahuja A, Fok TF, Poon WS. Cloverleaf skull-when should one operate? Singapore Med J 1997; 38: 217-220.
16. Heeckt P, Muhlbauer W, Fairley J, Anderl H, Hopner F. Pancraniofacial synostosis-indications for early craniofacial operation. Handchir Mikrochir Plast Chir 1994; 26: 60-67.
17. Carmel PW, Lukem MG, Ascherl GF. Craniosynostosis: Computed tomographic evulation of skull base and calvarial deformities and associated intracranial changes. Neurosurgery 1981; 9: 366-372.