

The sella turcica in children with lumbosacral myelomeningocele

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SUMMARY The purpose of the present study was to analyse the morphology of the sella turcica in children born with myelomeningocele.

Profile radiographs from 16 children (nine females and seven males) born with myelomeningocele were analysed.

The contour of the anterior wall of the sella turcica in myelomeningocele patients, instead of following the normal cranio-caudal direction, was always in an obliquely antero-posterior direction. The sella turcica thus appeared broad cranially with a diverging anterior wall, or with both diverging anterior and posterior walls. This appearance gave an impression of a wide sella turcica in myelomeningocele with less depth than normal.

The investigation has drawn attention to the fact that congenital malformations in the axial skeleton, even though, as in the case of myelomeningocele, they are located far from the cranial base, may have manifested themselves in the cranial base as well. The pathogenetic relationship between these manifestations is to be found in the early embryonic structure, the notochord. With the concept of embryological developmental fields, defined as areas with a common developmental origin, such as the notochordal field involved in myelomeningocele, new ways seem to be emerging for an improvement of aetiologically based diagnosis and treatment.

Introduction

Spina bifida is a congenital anomaly in which the neural tube has not closed during early fetal life. In most cases the malformation is situated lumbosacrally (Ingraham and Swan, 1943; Schwidde, 1952; Berry, 1992; Golden and Chernoff, 1995). The clinical consequences of a defect in neural tube closure range from none to fatal. In most cases of spina bifida only the vertebral arches are not closed. This can be seen in 15–20 per cent of healthy persons. In spina bifida cystica, a meningeal cyst protrudes from the vertebral canal in the affected region. In 10 per cent of these cases the cysts contain the dura and the arachnoidea only, termed meningocele. In 90 per cent of cases of spina bifida cystica the

neural tissue also protrudes, and the condition is called myelomeningocele.

The prevalence of myelomeningocele has been reported to be 1:1000 (Haslam, 1996), and according to Shapiro and Bartoshesky (1990) from 1:500 to 1:2000 live births. The children affected are most often treated surgically immediately after birth. In cases of myelomeningocele the nerve supply distal to the level of the defect is more or less destroyed, resulting in neurological symptoms from the lower limbs, bladder and bowel. Hydrocephalus, which most often requires shunting from the ventricles in the brain, is a condition often associated with myelomeningocele. At least 70 per cent of those with myelomeningocele have normal intelligence (Haslam, 1996).

In a recent study of the axial skeleton in human fetuses with myelomeningocele, malformation of the sella turcica was found in all cases, and in some cases malformation of the basilar part of the occipital bone was observed. Moreover, parts of the adenohypophyseal tissue were found localized on the external side of the cranial base in the pharyngeal mucosa (Kjær *et al.*, 1996).

The purpose of the present study was to clarify, against the background of the prenatal investigations, the morphology of the sella turcica in children born with myelomeningocele.

Subjects and methods

Profile radiographs from 16 children (nine females and seven males) born with myelomeningocele were analysed. These records were considered historical material representing existing profile radiographs in 12 cases from hospital records and in four cases from orthodontic records. The indications for the hospital profile radiographs were mainly control of ventricular shunts (10 children had ventricular shunts), while the profile radiographs were obtained for orthodontic treatment planning and cranial growth control.

The hospital material originated mainly from the National University Hospital (Rigshospitalet),

Copenhagen, and the orthodontic material from four different clinics in the municipal and county dental service system. Information concerning the myelomeningocele condition was available from hospital physicians and/or parents.

Drawings of the 16 sella turcicas were made and compared with earlier published standards for the morphological appearance of the sella turcica (Bergerhoff, 1963) and to standards, including the growth pattern from childhood to adulthood of the sella turcica, published by Björk and Skieller (1983).

Results

The distribution of the material according to the ages when radiographs were taken appears in Figure 1. The sella turcica area marked on the profile radiograph is shown in Figure 2 and examples of sella turcica morphology are given in Figures 3–6.

The drawings of the sella turcica contours from the radiographs of the 16 patients are included in Figure 1.

It is obvious from the drawings that the sella turcica morphology in myelomeningocele differs from the normal morphology, also shown in Figure 1. In all cases, the contour of the anterior wall of the sella turcica deviated from normal.

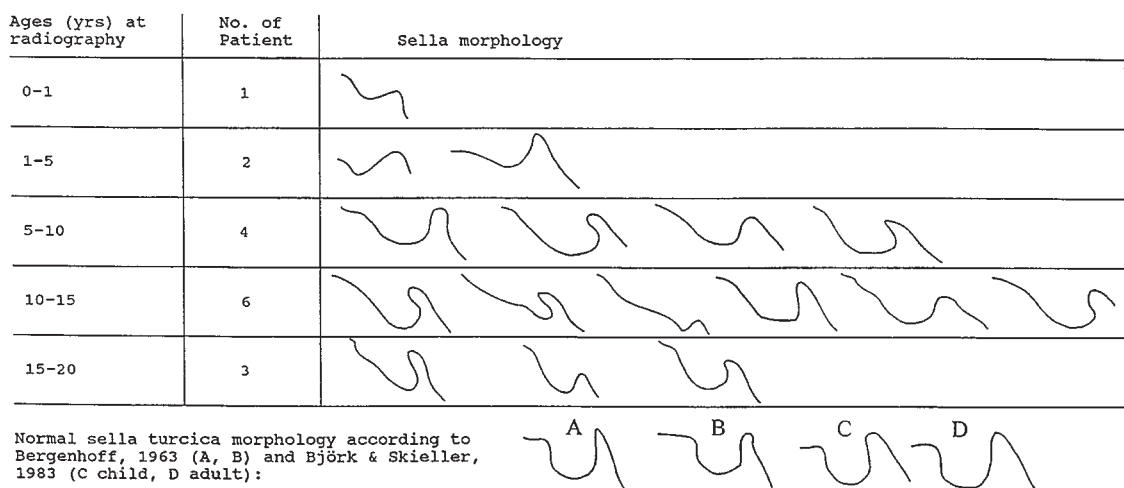


Figure 1 Sella turcica morphology in 16 patients with myelomeningocele. The anterior wall of the sella turcica (left in all drawings) is oblique in myelomeningocele, while the contour in normal development has a straight cranio-caudally orientated segment. This means that the sella turcica appears more open cranially in myelomeningocele.



Figure 2 Section of a lateral profile radiograph of the cranium of a girl, aged 8 years and 11 months, with myelomeningocele. Inset in the top right corner is a profile radiograph of a normally developed girl at the same stage of development. Two arrowheads indicate the sella turcica in the myelomeningocele patient and a single arrowhead marks the sella turcica in the normally developed girl. The anterior wall of the sella turcica is antero-posteriorly inclined in myelomeningocele and cranio-caudally orientated under normal conditions.

The contour of the anterior wall in myelomeningocele patients, instead of following a normal cranio-caudal direction, was always oblique with an antero-posterior orientation. The sella turcica thus appeared broad cranially with a diverging anterior wall, or with both diverging anterior and posterior walls. This appearance often resulted in an impression of a wide sella turcica in myelomeningocele with less depth than normal.

The morphological appearances seemed to be independent of age. The most deviant malformation (age group 10–15, No. 3, Figure 1) was seen in an institutionalized, severely mentally-retarded patient.



Figure 3 Lateral radiograph of the sella turcica from a girl with myelomeningocele, aged 2 years and 11 months. The anterior wall (left) is antero-posteriorly inclined and the posterior wall is displaced dorsally.

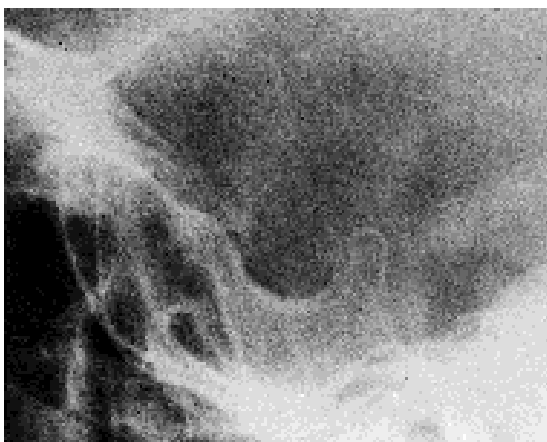


Figure 4 Lateral radiograph of the sella turcica from a girl with myelomeningocele, aged 7 years and 4 months. The anterior wall of the sella turcica (left) is antero-posteriorly inclined and the posterior wall is displaced dorsally.

Discussion

The malformations observed in the anterior wall of the sella turcica in patients with myelomeningocele have the same location as the malformations described prenatally in fetuses with this condition (Kjær *et al.*, 1996).

The malformations in the sella turcica of the patients are seen not only in the anterior wall of the sella, but in many cases also in the posterior

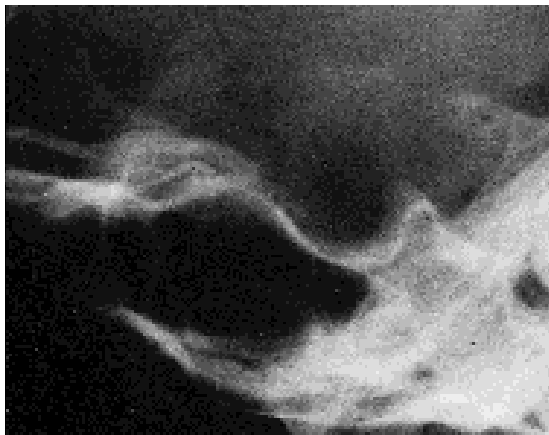


Figure 5 Lateral radiograph of the sella turcica from a boy with myelomeningocele, aged 8 years and 2 months. The anterior wall of the sella turcica (left) is antero-posteriorly inclined and the posterior wall is displaced dorsally.

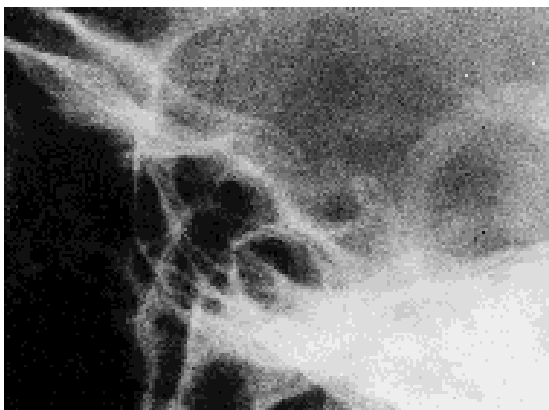


Figure 6 Lateral radiograph of the sella turcica from a boy with myelomeningocele, aged 13 years and 1 month. The anterior wall of the sella turcica (left) is antero-posteriorly inclined and the posterior wall has a normal appearance.

wall, leading to an impression of reduced depth of the sella turcica. Whether this change is due to the malformation as such, or to deformation caused by altered intracranial pressure related to the hydrocephalus condition or to the functioning of the shunt, cannot be determined (New, 1966; Kaufmann *et al.*, 1970; New and Weiner, 1971; Madsen, 1992).

The present study shows how the constant finding of a malformed sella turcica in prenatal

myelomeningocele is also found in postnatal myelomeningocele. The observation of a malformed sella turcica in patients with myelomeningocele is a finding which has not previously been reported.

In the prenatal myelomeningocele condition, malformation of the pituitary gland was also observed (Kjær *et al.*, 1996). The available patient records did not include data on endocrinological development, such as precocious puberty, which is often associated with brain lesions and myelomeningocele (Elias and Sadeghi-Nejad, 1994; DiGeorge and Garibaldi, 1996; Proos *et al.*, 1996). In relation to the prenatal finding of displaced extracranial adenohypophyseal tissue in cases with myelomeningocele (Kjær *et al.*, 1996) and reports on postnatal dysfunction in the hypothalamic-pituitary axes in basal encephaloceles (Lieblich *et al.*, 1978), it seems advisable to include such aspects in future studies of these patients.

In a wider perspective, prenatal accounts of the occurrence and extent of other malformations should form the basis for interdisciplinary studies of children with congenital malformations.

Studies of growth in the normal craniofacial skeleton have revealed that the contour of the perpendicular cranial segment of the anterior wall of the sella turcica is stable during growth in childhood (Björk and Skieller, 1983). This contour therefore serves as a reference structure for growth analysis when radiographs of the same patient taken at different ages are superimposed. Consequently, knowledge of sella turcica morphology is of great importance for orthodontic diagnosis and treatment. Because orthodontists regularly analyse a considerable number of profile radiographs, they will in many cases be the first to register minor malformations of the sella turcica. Insight into sella turcica malformations and information about the aetiological background of such malformations are not available in the literature. The present investigation is intended to be the first in a series of postnatal cranial base analyses, based on prenatal studies of different congenital malformations.

The present study has drawn attention to the fact that congenital malformations in the axial skeleton, even though, as in the case of

myelomeningocele, they are located far from the cranial base, may have manifested themselves in the cranial base as well. The pathogenetic relationship between these manifestations is to be found in the early embryonic structure, the notochord (Marín-Padilla, 1993), and in the interaction between the notochord and the surface ectoderm (Helwig *et al.*, 1995; Balling *et al.*, 1996). The notochord is responsible both for neural tube closure and for the development of the axial skeleton (Marín-Padilla, 1979, 1991; Müller and O'Rahilly, 1980; Saraga-Babic and Saraga, 1993; Kjær *et al.*, 1994). As the original course of the notochord extends from the coccygeal region to the sella turcica (Kjær, 1994, 1995), it is conceivable that dysfunction at the caudal end (lumbosacral myelomeningocele) can also be expected to occur at the cranial end (sella turcica and pituitary gland malformations) (Kjær and Fischer Hansen, 1995a,b).

With the concept of embryological developmental fields, defined as areas with a common developmental origin, such as the notochordal field involved in myelomeningocele, new ways seem to be emerging for an improvement of aetiologically based diagnostics.

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References

- Balling R *et al.* 1996 Pax genes and skeletal development. *Annals of the New York Academy of Sciences* 785: 27–33
- Bergerhoff W 1963 III Röntgenologische Schädelmesung. In: Diethelm L, Strnad F (eds) *Röntgendiagnostik des Schädels*. I. Springer-Verlag, Berlin pp. 102–105
- Berry C L 1992 A view of neurospinal dysraphism. *Virchows Archiv (A)* 420: 375–376
- Björk A, Skieller V 1983 Normal and abnormal growth of the mandible. A synthesis of longitudinal cephalometric implant studies over a period of 25 years. *European Journal of Orthodontics* 5: 1–46
- DiGeorge A M, Garibaldi L 1996 Disorders of pubertal development. In: Behrman R E, Kliegman R M, Arvin A M (eds) *Nelson textbook of pediatrics*, 15th edn. W B Saunders Company, Philadelphia, pp. 1580–1583
- Elias E R, Sadeghi-Nejad A 1994 Precocious puberty in girls with myelodysplasia. *Pediatrics* 93: 521–522
- Golden J A, Chernoff G F 1995 Multiple sites of anterior neural tube closure in humans: evidence from anterior neural tube defects (anencephaly). *Pediatrics* 95: 506–510
- Haslam R H A 1996 The nervous system/neurologic evaluation. In: Behrman R E, Kliegman R M, Arvin A M (eds) *Nelson textbook of pediatrics*, 15th edn. W B Saunders Company, Philadelphia, pp. 1667–1679
- Helwig H *et al.* 1995 Interaction between *undulated* and *Patch* leads to an extreme form of spina bifida in double-mutant mice. *Nature Genetics* 11: 60–63
- Ingraham F D, Swan H 1943 Spina bifida and cranium bifidum. *New England Journal of Medicine* 228: 559–563
- Kaufmann M, Sandstrom P H, Young H F 1970 Alteration in size and configuration of the sella turcica as the result of prolonged cerebrospinal fluid shunting. *Radiology* 97: 537–542
- Kjær I 1994 The prenatal axial skeleton as marker of normal and pathological development of the human central nervous system. In: Lou H, Greisen G, Falck Larsen J (eds) *Alfred Benzon Symposium: brain lesions in the newborn*. Munksgaard, Copenhagen 37: 124–132
- Kjær I 1995 Human prenatal craniofacial development related to brain development under normal and pathologic conditions. *Acta Odontologica Scandinavica* 53: 135–143
- Kjær I, Keeling J W, Græm N 1994 Cranial base and vertebral column in human anencephalic fetuses. *Journal of Craniofacial Genetics and Developmental Biology* 14: 235–244
- Kjær I, Fischer-Hansen B 1995a The adenohypophysis and the cranial base in early human development. *Journal of Craniofacial Genetics and Developmental Biology* 15: 157–161
- Kjær I, Fischer-Hansen B 1995b Human fetal pituitary gland in holoprosencephaly and anencephaly. *Journal of Craniofacial Genetics and Developmental Biology* 15: 222–229
- Kjær I, Fischer Hansen B, Keeling J W 1996 Axial skeleton and pituitary gland in human fetuses with spina bifida and cranial encephalocele. *Pediatric Pathology* 16: 909–926
- Lieblich J M, Rosen S W, Guyda H, Rearden J, Schaaf M 1978 The syndrome of basal encephalocele and hypothalamic-pituitary dysfunction. *Annals of Internal Medicine* 89: 910–916

- Madsen P H S 1992 Kraniel morfologi og kraniel vækst hos børn behandlet for hydrocephalus med ventrikulær shunt. Thesis, University of Copenhagen, Denmark
- Marín-Padilla M 1979 Notochordal-basichondrocranium relationships: Abnormalities in experimental axial skeletal (dysraphic) disorders. *Journal of Embryology and Experimental Morphology* 53: 15–38
- Marín-Padilla M 1991 Cephalic axial skeletal-neural dysraphic disorders: Embryology and pathology. *Canadian Journal of Neurological Sciences* 18: 153–169
- Marín-Padilla M 1993 Notochordal alterations in axial skeletal-neural disorders. *Virchows Archiv (A)* 422: 97–98
- Müller F, O'Rahilly R 1980 The human chondrocranium at the end of the embryonic period, proper, with particular reference to the nervous system. *American Journal of Anatomy* 159: 33–58
- New P F J 1966 The sella turcica as a mirror of disease. *Radiologic Clinics of North America* 4: 75–92
- New P F J, Weiner M A 1971 The radiological investigation of hydrocephalus. *Radiologic Clinics of North America* 9: 117–140
- Proos L A, Dahl M, Ahlsten G, Tuvemo T, Gustafsson J 1996 Increased perinatal intracranial pressure and prediction of early puberty in girls with myelomeningocele. *Archives of Diseases in Childhood* 75: 42–45
- Saraga-Babic M, Saraga M 1993 Role of the notochord in the development of cephalic structures in normal and anencephalic human fetuses. *Virchows Archiv (A)* 422: 161–168
- Schwidde J T 1952 Spina bifida. *American Journal of Diseases of Children* 84: 35–51
- Shapiro K, Bartoshesky L E 1990 Meningomyelocele In: Buyse M L (ed.) *Birth defects encyclopedia*. Vol. II. Blackwell Scientific Publications Inc., Cambridge, MA, pp. 1120–1121

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