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Interhemispheric lipoma with variable callosal dysgenesis: relationship between embryology, morphology, and symptomatology

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Abstract. Eight interhemispheric lipomas (five tubulonodular lipomas and three curvilinear lipomas) were examined by magnetic resonance imaging (MRI). The purpose was to further investigate the relationship between the morphology of the different subtypes and the clinical presentation. The imaging findings were reviewed in light of a recent theory on the development of the corpus callosum. Interhemispheric lipomas should be considered as one entity with a variable expression depending on the severity and/or the time of the insult. Curvilinear lipomas can be either small or extensive and are usually not symptomatic. Tubulonodular lipomas can be either predominantly anterior or posterior in location. The anterior subtype appears to be a more severe form of tubulonodular lipoma. The associated structural abnormalities are most likely responsible for the symptoms, rather than the lipoma itself. Magnetic resonance imaging allows a more precise timing of the insult, resulting in the development of a lipoma. The knowledge of the embryology between the 6th and the 20th week is important to explain these abnormalities. Until now it has been accepted that the corpus callosum develops in an orderly fashion. A recent theory has demonstrated that this is not necessarily true, and that fibers can cross the midline at any place irrespective of the normal development. This theory explains the sometimes amorphous appearance of the remnant of the corpus callosum if a lipoma is present.

Key words: Corpus callosum – Lipoma and lipomatosis

Introduction

Interhemispheric lipomas, often misnamed corpus callosum lipomas, are uncommon congenital malformations occurring with an autopsy frequency of 1/2500 to 1/25 000 cases [1]. The origin of intracranial lipomas lies in a maldifferentiation of the primitive meninx which, instead of resorbing, persists and develops into mature adipose tissue. This theory was suggested by Verga in 1929 and is still accepted as the most likely to be true [2]. Truwit and Barkovich distinguished two subgroups of interhemispheric lipomas: the curvilinear (CL) lipoma and the tubulonodular (TN) lipoma [3]. They concluded that the latter must result from a more severe insult occurring at an earlier age and therefore interfering with the normal development of the corpus callosum. The corpus callosum is characterized by a very orderly development occurring between the 11th and 20th week of gestation, and it is generally accepted that the timing of an in utero injury is reflected in its morphology. More recently, Rubinstein et al. suggested that callosal dysgenesis may be caused by arrested growth or by delayed continued development in an attempt to compensate for previous midline abnormalities [4]. In light of these findings, we reviewed our imaging findings in eight lipomas. The purpose was to further try to elucidate the embryopathology of these congenital midline abnormalities and to correlate the clinical and radiological findings.

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Embryology

In assessing interhemispheric lipomas and associated brain abnormalities, it is important to know the development of the commissures and the septum pellucidum, on one hand, and the resorption of the meninx primitiva, on the other hand [5].

At 6–8 weeks the cranial end of the neural tube develops into a thin ventral part, the lamina terminalis, and a thick dorsal part, the lamina reuniens. The ventral part of the lamina reuniens becomes the anterior commissure (10 weeks). At 8–9 weeks the sulcus medianus telencephali medii develops from a median unfolding of the lamina reuniens. Both sides of this sulcus approximate and are separated only by a single mesenchymal

Table 1. Eight patients with an interhemispheric lipoma. TN tubulonodular; p posterior location; a anterior location; CL curvilinear

Patient no.	Age (years)	Gender	Morphology	Signs and symptoms	Final diagnosis
1	45	F	CL	Cervicobrachialgia	Cervical arthrosis
2	58	M	CL	Diplopia	Bilateral 6th nerve paresis
3	20	F	CL	Visual disturbances	Craniopharyngioma
4	8	M	TN, p	Seizures, dysmorphic facies	Seizures
5	18	M	TN, a	Seizures, dysmorphic facies, mild psychomotor retardation	Seizures
6	28	M	TN, p	Recent complex partial seizures	Seizures
7	33	M	TN, p	Dysmorphic facies	No diagnosis
8	53	F	TN, p	Bilateral pyramidal signs	Meningioma in the cervical spinal canal

fold of the meninx primitiva (10 weeks). The massa commissuralis is formed when the fusion of the sulcus occurs. This is the predecessor of the corpus callosum. The corpus callosum develops between 11 and 20 weeks. The genu is formed between 14 and 16 weeks, the truncus between 15 and 20 weeks, the splenium between 17 and 19 weeks, and the rostrum between 19 and 20 weeks. From 20 weeks until birth the fully formed corpus callosum increases in size, but remains morphologically identical.

The meninx primitiva forms the subarachnoid space. At 8 weeks the meninx has resorbed completely, except for a small quantity located dorsal to the lamina terminalis.

The fornices and the hippocampal commissure develop from the dorsal part of the lamina reuniens (9–11 weeks). The development of the septum pellucidum and the cavum septi are closely related to the formation of the massa commissuralis.

The sulcus telencephali medii, filled with the meninx primitiva, remains open to the interhemispheric fissure in its ventral part (12–13 weeks). With the development of the corpus callosum, a diastase occurs in the base of the sulcus and a subcallosal pocket is formed: the cavum septi pellucidi. The cavum is separated anteriorly from the interhemispheric fissure by the fibers of the rostrum of the corpus callosum (19–20 weeks). The cavum closes after birth as the white matter of the cerebral hemispheres increases. The septum pellucidum is a small membrane extending from the inferior border of the corpus callosum to the fornix.

The choroid plexus is formed by an invagination of the ependyma and the pia along the medial wall of the hemispheres (or telencephalic vesicles) into the lateral ventricles and third ventricle (7–10 weeks).

Patients and methods

Eight patients (five males and three females) with an interhemispheric lipoma were examined by MRI. The patients ranged in age from 8 to 58 years with an average age of 33 years (Table 1).

Five patients had a TN lipoma and three patients had a CL lipoma. All MRI examinations were performed at 1.5 T. Sagittal and/or coronal spin-echo (SE) T1-weighted images were obtained in eight and six patients, respectively. Axial T1- and T2-weighted images were obtained in three and four patients, respectively. Cranial CT was available in all patients. The lipomas were classified as either CL or TN based on the morphological criteria in the literature [3].

The following imaging features were studied: morphology of the corpus callosum, the fornix, the septum pellucidum, and the anterior commissure; distribution of the lipomatous tissue (pericallosal, interhemispheric, in the septum pellucidum, in the choroid plexus, etc.); location of the pericallosal/callosomarginal artery, presence of calcification on CT, calloso-sellar distance (distance, perpendicular to a horizontal line, between the inferior border of the corpus callosum and the dorsum sellae on the midline sagittal slice through the aqueduct). Based on this perpendicular line, we decided on a predominantly anterior or posterior location of the TN lipoma depending on where most lipomatous tissue was seen.

Results

Three patients with a curvilinear (CL) lipoma and five patients with a tubulonodular (TN) lipoma were examined by MRI. The corpus callosum was entirely normal in the three patients with a CL lipoma (Fig. 1). The maximal width of the lipoma on the axial images never exceeded 1 cm. There was no calcification and the pericallosal artery was always identified cranial to the lipoma. There was a small left-sided choroid plexus lipoma in one of three patients. The anterior commissure was normal in one patient, small in the second, and could not be identified in the third, due to the presence of a suprasellar craniopharyngioma. The fornix was normal in all three patients, but both legs were asymmetrical in one patient. A small cavum septi pellucidi was seen in one patient and the septum pellucidum was normal in the other two patients. All three patients had symptoms, but these were not related to the presence of the lipoma. The calloso-sellar distance varied between 3.5 and 4.6 cm (mean 4.1 cm).

The corpus callosum was abnormal in all five patients with a TN subtype. The bulk of the lipoma was located

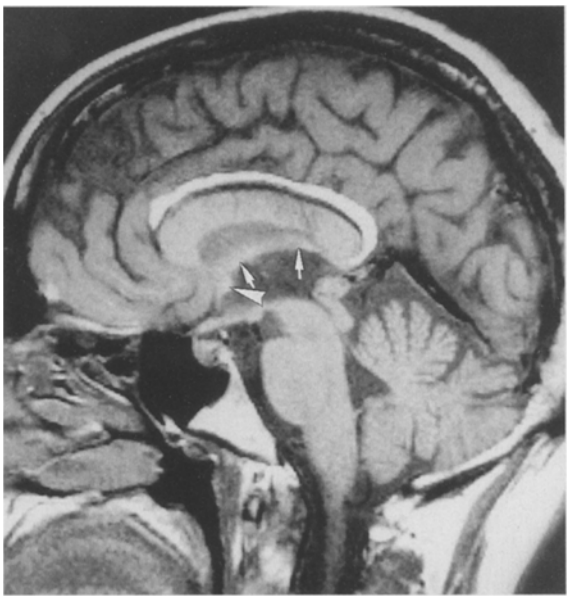


Fig. 1. Sagittal T1 weighted (TR/TE 600/15 ms) image (case 1). A typical curvilinear interhemispheric lipoma is shown. Note the normal morphology of the corpus callosum, fornix *(arrow)*, and anterior commissure *(arrowhead)*

Fig. 2 a–b. Sagittal and coronal T1-weighted (TR/TE 520/15 ms) images [case 5 **(a, b)**]. An anterior tubulonodular interhemispheric lipoma with a large posterior component. Note the normal anterior commissure (**a**; *arrowhead*). The fornix appears dysgenetic and cannot be separated from the part of the corpus callosum that has developed. An extension of the lipoma in the interhemispheric fissure is present (**a**; *arrow*). Lipomatous components are seen in the septum pellucidum and in the left choroid plexus (**b;** *arrow*)

Fig. 3 a–d. Sagittal and coronal T1-weighted (TR/TE 520/15 ms) images (case 4 **a,** case 7 **b, c,** and case 8 **d**). Three different posterior tubulonodular interhemispheric lipomas are shown. Note the anterior peg-like extension of the lipomas. The anterior commissure appears normal *(arrow),* except in **b** where the anterior commissure appears hypoplastic. Note the bilateral extension of the lipoma in the choroid plexus in **c**. The genu of the corpus callosum is seen in all three patients, but the truncus is only partly present in **b** and **d**

anteriorly in one of five patients, but an important posterior component was present (Fig. 2). The posteriorly located TN lipomas in four of five patients were characterized by an anterior peg-like extension of the lipomatous tissue (Fig. 3). The maximal width in the axial plane always exceeded 1 cm. Calcification was present in two of five patients on CT. The pericallosal artery ran through the lipoma in four of five patients. Extension of the lipoma into the choroid plexus was seen in four of five patients, in two of them, bilaterally. There was evidence of lipomatous tissue in the septum pellucidum in 1 patient and in the anterior left lateral ventricle in another patient. The anterior commissure was normal in four of five patients and was small in the remaining patient. The fornix could not be separated from the corpus callosum in three of five patients and was small in two of five patients. There was a cavum vergae in two patients. The callososellar distance varied between 2.7 and 4.0 cm (mean 3.2 cm).

The symptoms in the three patients with a CL lipoma were unrelated to the lipoma. One patient complained of cervicobrachialgia, one patient had bilateral sixth nerve paresis, and another patient had a large suprasellar craniopharyngioma.

In the group of patients with a TN lipoma, seizures and a dysmorphic facies were present in two of five patients. One patient had complex partial seizures and one patient only had a dysmorphic facies. The fifth patient had bilateral pyramidal signs and an intraspinal meningioma at the cervical level. The results are summarized in Table 2.

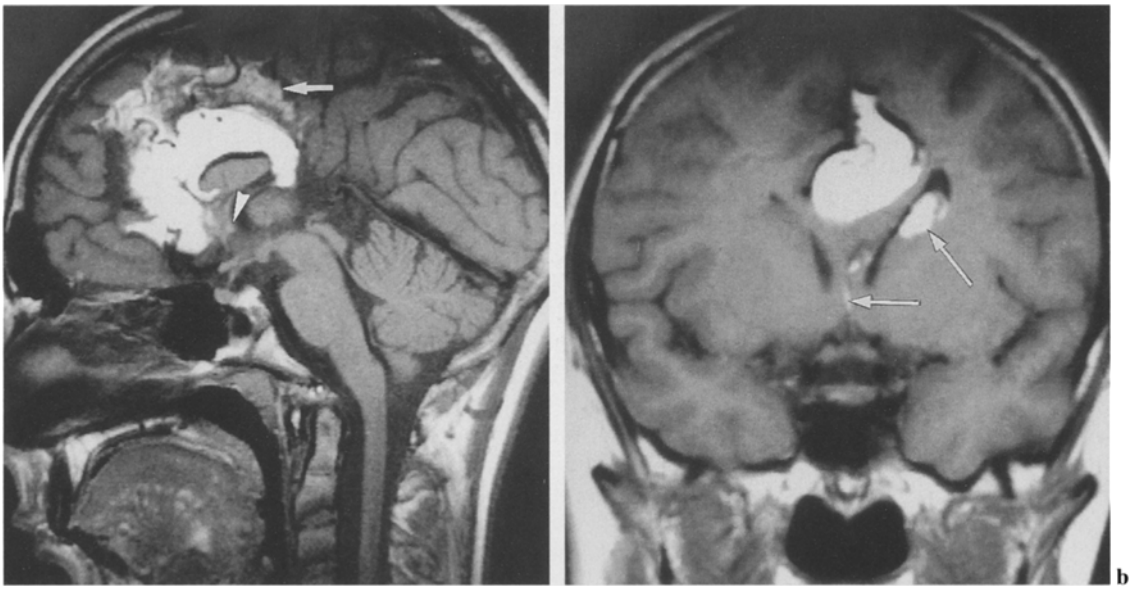
Discussion

Interhemispheric lipomas are rare congenital malformations. This abnormality was first described by Rokitansky in 1856 [6]. The concept of a persistent focus of primitive meninx that develops into mature adipose tissue was suggested by Verga in 1929 [2]. With the advent of MRI, several authors have reported small series of interhemispheric lipomas. Truwit and Barkovich have sug-

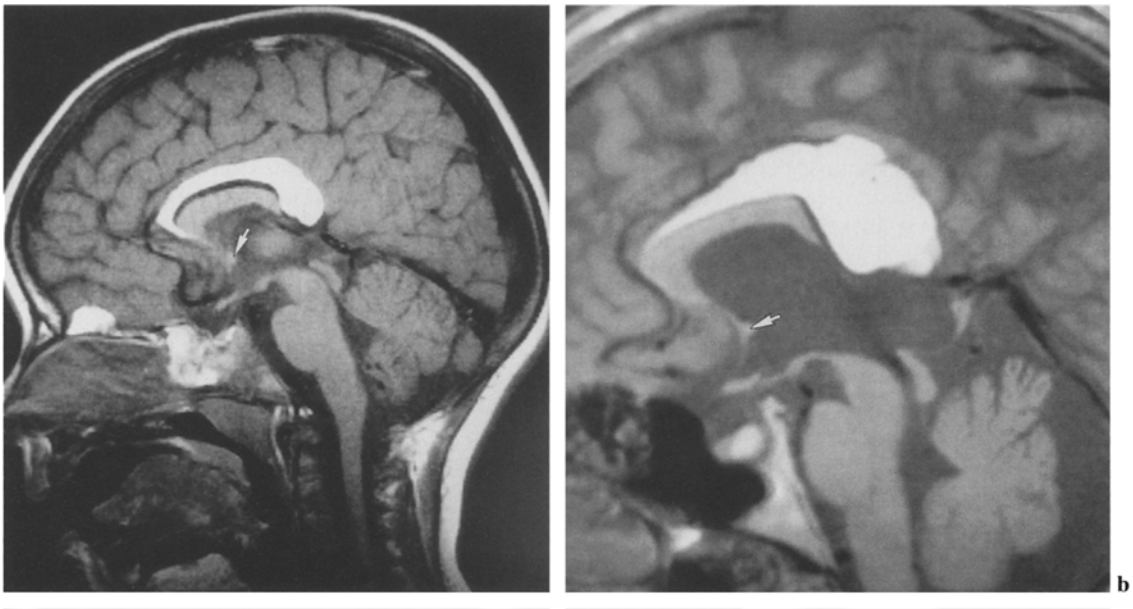
Table 2. MRI findings in eight patients with an interhemispheric lipoma. + present; – absent; ± partly present

Patient no.	Morphology of corpus callosum				Fornix	Septum pellucidum	Callosal distance (cm)[a]	Pericalosal artery location with respect to lipoma	Anterior commissure	Presence of lipoma in choroid plexus		Ca^{2+} on CT
	Rostrum	Genu	Truncus	Splenium						Left	Right	
1	+	+	+	+	Asymmetry of both legs	Normal	3.5	Above	Normal	–	–	–
2	+	+	+	+	Normal	Cavum septi pellucidi	4.2	Above	Normal	+	–	–
3	+	+	+	+	Normal	Normal	4.6	Above	Normal	–	–	–
4	–	+	+	–	Dysgenesis	Normal	3.8	? Above	Normal	+	–	–
5	–	–	±	–	Dysgenesis	Lipoma	3.1	In	Normal	+	–	+
6	–	+	±	–	Dysgenesis	Not visualized	2.5	In	Normal	+	+	–
7	–	+	±	–	Hypoplastic	Cavum vergae	4.0	In	Hypoplastic	+	+	+
8	–	+	±	–	Hypoplastic	Cavum vergae	2.7	In	Normal	–	–	–

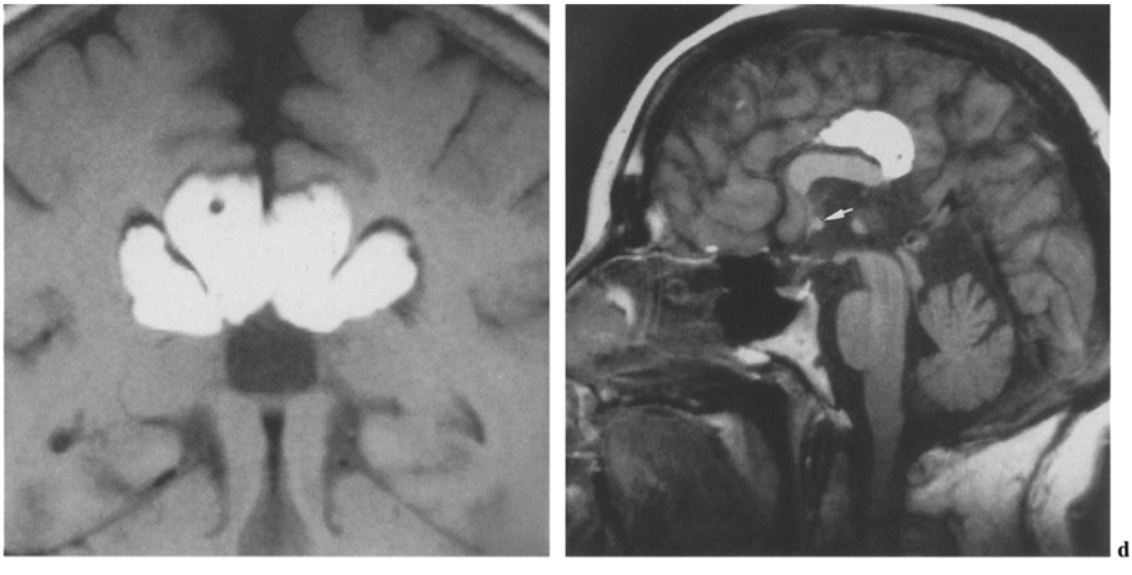
[a] Distance between inferior border of corpus callosum and dorsum sellae on midline slice through aqueduct



2a b



3a b



c d

gested that these lipomas may have different morphological appearances according to the time and/or severity of the insult [3]. They introduced the CL and the TN subtype, the latter being associated with other congenital abnormalities [7].

Recently, it has been demonstrated that callosal dysgenesis may also be the result of delayed continued development after an insult [4]. This observation is important in the study of interhemispheric lipomas. Until recently, it was accepted that when the commissural plate fails to form or is damaged, the callosal fiber cannot cross the midline. The new theory demonstrates that fibers are able to cross the midline at any place. The usual sequence of formation of the genu, truncus, splenium, and rostrum is therefore not always respected.

We reviewed eight pericallosal lipomas in light of these recent findings. The CL lipomas were never associated with corpus callosum abnormalities and the symptoms were not related to the presence of the lipoma.

All five TN lipomas were associated with callosal dysgenesis as well as with other anatomical abnormalities. The associated dysmorphic facies in three of five patients with a TN lipoma was probably related to the presence of the interhemispheric lipoma. The seizures in three of five patients can also be due to the intracranial morphological abnormalities. Interhemispheric lipomas are associated with a significant seizure disorder in 50 % of cases. The hypothesis to explain the seizures remains controversial. It may result from the hemispheric disconnection, but an infiltration of the surrounding brain tissue by the fibrous capsule of the lipoma is another possibility [1, 8].

The primitive meninx is completely resorbed by the 10th week. At that time the massa commissuralis is barely formed and the corpus callosum develops only between the 11th and 20th week [5]. Therefore, all interhemispheric lipomas must arise from remnants of the primitive meninx. The earlier and/or more severe the insult is, the more important the abnormalities. It is suggested that the CL lipomas originate later in pregnancy and/or result from a minor insult and do not interfere with the development of the corpus callosum.

The TN lipomas originate at some time between the 12th and the 20th week and/or result from a more severe insult that leads to callosal dysgenesis. The insult also affects other structures that are developing at that time such as the fornix and the septum pellucidum. The fornix was abnormal in three of five patients and a cavum vergae was seen in two and five patients with a TN subtype. The anterior commissure develops from the ventral part of the lamina reuniens and was normal in all cases. The pericallosal artery is present by the 10th week of gestation and the TN lipoma therefore most likely surrounds the artery. In patients with a CL lipoma the artery was always localized cranially to the lipoma. Associated choroid plexus lipomas were seen in four of five patients, always in contiguity with the interhemispheric lipoma. The frequency of extension of the interhemispheric lipoma in the choroid plexus has been 20–25 % of the cases in the literature [9]. In our limited series the frequency was 33 % in the group of CL lipomas and 80 % in the group of TN lipomas. This is not surprising and supports the hypothesis that TN lipomas are the result of a previous and/or more severe insult, because it is known that the choroid plexus is formed very early in development (7–10 weeks).

In one patient there was involvement of the choroid plexus in the left anterior lateral ventricle, which is an extremely unusual finding [10]. A lipomatous component in the septum pellucidum was seen in one patient. In another patient the structure of the abnormal corpus callosum was not interpretable. In this patient only a small part of the corpus callosum was seen, most likely a part of the truncus. This can be explained by the theory of Rubinstein et al. [4]. The truncus should then be considered as the only location where fibers were able to cross the midline.

There were no associated frontal bone defects or encephaloceles in our series, but these findings have been observed in more than 40 % of the TN lipomas in the literature [7].

We suggest to use the term interhemispheric lipoma, because the degree of callosal dysgenesis can vary significantly and sometimes the anatomy of the corpus callosum is not recognizable. Therefore, terms such as pericallosal and corpus callosum lipoma, are less accurate.

Until now, only so-called anterior TN lipomas have been reported. We also found posterior TN lipomas which were always associated with an anterior peg-like extension.

We suggest that interhemispheric lipomas should be considered as one entity with a variable expression depending on the time and/or severity of the insult. The TN subtype most likely is the result of a more severe or previous insult. The TN lipomas are commonly associated with other anatomical abnormalities in structures that are developing at the time of insult. From our limited experience we suggest that the anterior TN lipoma is the result of a more severe or earlier insult than the posterior TN lipoma.

A clinicoradiological correlation reveals that CL lipomas do not cause symptoms and that the symptoms in TN lipomas most likely result from the associated abnormalities, rather than from the lipoma itself.

Our findings illustrate the theory recently introduced by Rubinstein et al. [4]. The presence of an isolated part of the corpus callosum, a finding that cannot embryologically be explained if an orderly development is accepted, indicates that callosal fibers can cross the midline after an insult if they find a passage. Magnetic resonance imaging is not necessary for the diagnosis of a lipoma, but it allows a more precise analysis of the associated abnormalities and therefore may allow a more precise timing of the insult, an important contribution in medicolegal cases. Furthermore, the assessment of the associated abnormalities is best achieved on MRI images.

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Book review

European Radiology

Castillo M., Mukherji S.K.: Imaging of the Pediatric Head, Neck and Spine. Philadelphia, New York: Lippincott-Raven Publishers 1996, 768 pages, 1110 illustrations, 33 tables, $ 171.50,–. ISBN 0-397-51577-4

Books aiming at a comprehensive representation of the total field of neuroradiology have become seldom due to the rapid development and increase in knowledge. Nevertheless, Castillo and Mukherji have undertaken the attempt in paediatric neuroradiology. The authors, well known by many publications, try to give concentrated information in three chapters (brain, head and neck, spine) and they succeed brilliantly. They have written a book for everyday use with a description of practically all – even the most seldom – diseases, and a clear structure.

The first chapter of this logically organized book gives a short methodology on techniques, sequences and methods used by the authors up to sedation and anaesthesia. It is shown again that clinical symptoms and supposed diagnosis are of absolutely first priority for the authors. The examination protocols are rigorously adapted to clinical findings and stress the clinical character of an efficient paediatric neuroradiology. Modern equipment is used specifically and not for screening purposes.

Although trying to give maximal information in minimal space, the authors do not omit important basics, such as embryology, which is found at the beginning of each chapter dealing with developmental anomalies. In this section all anomalies, even the most (unusual) eccentric ones, are discussed and demonstrated by images of good quality. The presentation of each entity follows the same line: "imaging features" being discussed only after "epidemiologic data" and "clinical features".

The chapter "Metabolic and Degenerative Disorders" presents good comprehensive and comparative tables, useful and clear for differential diagnosis. It is shown especially by this chapter that the book deals with the full range of neuroradiology with all its refinements in technique and methods. The disorders are also discussed with respect to MR spectroscopy.

Diseases frequently seen by the radiologist in practice as vascular processes or tumours are presented in detail with the same accuracy. Rare entities, such as, for example, "dysembryoplastic neuroepithelial tumours" are to be found as well, also by imaging. Midline venous thrombosis, a rare disorder unfortunately occasionally diagnosed by less-experienced examiners, is found in this book as well, illustrated by typical images. Attention is paid to the frequent forms of hydrocephalus. Neurocutaneous syndromes are especially well described and summed up in a table to facilitate differential diagnosis.

The second main chapter, "Head and Neck" is an especially well-done part of the book. For better understanding all malformations are introduced by "applied embryology". As in the whole book, all possible, and for the diagnosis of the disorders suitable neuroradiological methods, are presented here, as for instance 3D reconstruction of midline clefts and agenesis of facial structures. Radiological images are supplemented by very impressive photographs of the patients emphasizing the clinical orientation of the book. Examinations of high quality are demonstrated in the section "Temporal bone". All anomalies are precisely described and accompanied by respective examples. Marking of the pathologic signs is clear, demonstrative and not overloaded.

Disorders and pathologic findings of "Neck" are fully and precisely specified. Here again it is obvious that the section "clinical features" is clearly more detailed than the actual radiologic "imaging features" themselves. The importance of information on clinical findings is emphasized again.

The last chapter deals with "Spine". Malformations of upper spinal level – highly significant for paediatric neurosurgery – are described first in the known order and are well demonstrated by good images. The same applies to the chapter on inflammations and tumours.

Other publications are rarely cited, stating the broad knowledge and the great personal experience of the authors. Nevertheless, they often refer to important larger publications, mostly review articles ("Suggested readings").

On the whole, the book can be regarded as a very successful and detailed, but also concentrated and precise, up-to-date publication. Imaging material is of high quality, important for every radiologist and commensurate with modern times and equipment. This applies even to rare entities. Fortunately, findings are not based on a single method alone, e.g. MRI. The book does not concentrate on the method but on the organ of interest. All known diseases of the (respective region) region concerned are taken into account. The neuroradiologist alone would like to see more angiographic findings in the chapters concerned. The authors, however, cannot be blamed for this deficiency with the book aimed not only at neuroradiologists and made for a wide everyday use; besides, more angiograms would certainly have exceeded the planned volume. In conclusion, this manual of this broad field is of high and mostly actual standard, apart from interventional neuroradiology. It should be on the bookshelf of everyone working in the radiological, clinical or surgical field.

L. Solymosi, Würzburg