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Congenital lumbosacral lipomas

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Abstract Congenital lumbosacral lipomas can be responsible for progressive defects. The general feeling is that tethering of roots, filum, or cord probably explains this evolution, and that untethering of these structures could prevent late deterioration. Like the vast majority of neurosurgeons, we too have routinely and systematically operated on lumbosacral lipomas, even in the absence of neurological deficits. This policy stemmed from our belief that spontaneous neurological deterioration was frequent, recovery from preoperative deficits rare, and surgery both efficient and benign in nature. After 22 years of experience, we felt that it was necessary to review our series of 291 lipomas (38 lipomas of the filum and 253 of the conus) operated on from 1972 to 1994. To reassess the value of prophylactic surgery, we attempted an accurate evaluation of (1) the risk of pathology, (2) the risks involved in surgery, (3) the postoperative outcome with respect to preoperative deficits, and (4) the postoperative outcome in asymptomatic patients at 1 year and at maximum follow-up. Special attention was paid to 93 patients whose postoperative follow-up was more than 5 years (average 8.7, median 8, range 5–23 years). Of these 93 patients, 39 were asymptomatic preoperatively (7 with lipoma of the filum and 32 with lipoma of the conus). Lipomas of the filum and of the conus are entirely different lesions and were studied separately. In 6 cases prenatal diagnosis had been possible. The mean age at surgery was 6.4 years. Low back skin stigmata were present in 89.4% of cases. Preoperative neurological deficits existed in 57% of the patients and were congenital in 22%. Clinical signs and symptoms recorded were pain in 13.3% of the patients and/or neurological deficits affecting sphincter (52%), motor (27.6%) and sensory (22.4%) functions. Deficits were progressive in 22.4% of cases, slowly progressive in 58.8% of these and rapidly progressive in the remaining 41.2%. In 36 patients (13.2%) the lipomas were seen to grow either subcutaneously or intraspinally. Among these patients, 21 were infants, 2 were obese adolescents, and 10 were pregnant women. The metabolism of the fat within the lipomas was studied in 11 patients and found to be similar to that at other sites. Lipomas were associated with various other malformations, either intra- or extraspinal. These associated anomalies were rare in the case of lipomatous filum (5.2%) but frequent with lipomas of the conus, except for intracranial malformations (3.6%). Therapeutic objectives were spinal cord untethering and decompression, sparing of functional neural tissue and prevention of retethering. Procedures used to achieve these goals were subtotal removal of the lipoma, intraoperative monitoring, duroplasty, and some-

times closure of the placode. Histologically, lipomas consisted of normal mature fat. However, 77% of them also included a wide variety of other tissues, originating from ectoderm, mesoderm, or entoderm. This indicates that lipomas are either simple or complex teratomas. The results of the study are as follows. (1) Surgery was easy and safe when performed for treatment of lipomas of the filum (no complications), but difficult and hazardous in the case of lipomas of the conus (20% local, 3.9% neurological complications). (2) All types of deficit could be improved by surgery, which was beneficial in all cases of lipoma of the filum and 50% of cases of lipoma of the conus. (3) In asymptomatic patients long-term surgical results depended on the anatomical type of the lipoma. They were excellent in lipomas of the filum. In lipomas of the conus they were good in the short term but eroded with time. At more than 5 years of follow-up only 53.1% of the patients were still free of symptoms. (4) Reoperations were performed in 16 patients (5.5%), 5 (31.2%) of whom improved postoperatively, while in 7 (43.7%) progression stopped, in 3 (18.7%) deterioration continued and in 1 (6.2%) the condition was worse after surgery. (5) The natural history of the malformation, that is to say the risk of spontaneous aggravation, has only been evaluated in hospital inpatients, so that the true level of risk remains unknown. This means we cannot interpret the actuarial curve following surgery for asymptomatic lipoma of the conus. In conclusion, there are two different types of lipoma: lipoma of the filum, for which surgery is harmless and beneficial in both the short and the long term, and lipoma of the conus, for which surgery involves considerable risks and is of questionable benefit in the long term. This raises the question as to whether prophylactic surgery is indicated for patients with asymptomatic lipomas of the conus, and whether the outcome is any better than it would be if the lipoma were left to take its natural course. The lack of basic information remains a stumbling block to management of these patients. Until this is remedied, we are unable to recommend prophylactic surgery in patients with asymptomatic lipomas of the conus.

Key words Lumbosacral lipoma · Spina bifida · Prophylactic surgery · Prenatal diagnosis · Tethered cord

Introduction

Congenital lumbosacral lipomas have been extensively studied. Since their first description, more than 2000 cases have been reported in the French and English literature, most of them in the past 50 years. Even at the end of the last century, their anatomy had already been almost fully described [67, 173, 182, 190]. By 1916, their clinical presentation was well known, and risks of spontaneous neurological deterioration were widely emphasized [174]. Tethering of roots [174], filum [91], or cord [111] was described in early reports as a frequent accompanying feature and believed to be a major cause of neurological deterioration. As a consequence, untethering of cord and roots progressively emerged as a possible way of preventing further neurological deterioration. In 1918, Brickner [29] operated on a symptom-free patient. In 1937, Leveuf [115] questioned whether early surgery could avoid the later development of deficits. In 1950, Basset [18] strongly advocated prophylactic surgery. At present, most neurosurgeons advocate early and preventive removal of lipomas, mainly for rapid untethering of the nervous system [7, 11, 12, 19, 32, 45, 47, 71, 75, 79, 84, 93, 94, 109, 111, 116, 117, 119, 126, 145, 153, 157, 175, 184, 189, 193, 196]. This has also been our policy [147, 148]. Since 1972, 291 of our 303 patients with a lumbosacral lipoma have been operated upon, the vast majority following first presentation, regardless of their clinical condition. Routine and prophylactic surgery was justified by our strong feeling that the risk of spontaneous neurological deterioration was high, the probability of postoperative regression of preoperative deficits was low, and surgery was benign in nature and offered the best chance of prevention.

If this is indeed the case, why bring up the subject for reconsideration? There are two reasons. First, many aspects of lipomas remain poorly understood or unknown as yet. This is the case for their embryogenesis, nature, and rate of incidence. Second, there is no universal agreement on the management of these, as may appear at first sight. The small number of publications suggesting that surgery should not be performed routinely [23, 28, 106, 152, 178] or favoring diverse attitudes according to patient's age [47, 101] does not reflect reality. In fact, many neurosurgeons and pediatricians are still reluctant to consider routine surgery in asymptomatic patients. Besides the problem of proposing surgery in the absence of deficits, the arguments that have been put forward include our continuing ignorance of the natural evolution of lipomas, which has been evaluated only in patients who have been operated upon, the wide variation in operative results among series, and the very short postoperative follow-up reported in most series.

All these remarks are relevant. Healthy adult carriers of the malformation are never included in medical statistics; their frequency is, indeed, unknown. Postoperative follow-

up is not mentioned in 72% of the publications [149] and, when published, does not exceed an average of 2.5 years. Finally, only two series, each with a maximum of 10 cases, compare the course in operated and non-operated patients [32, 106].

The value of routine and preventive surgery must therefore be reappraised. This implies weighing and comparing the risks of surgery against those of the disease itself, as well as assessing the efficiency of surgery in the short and in the long term.

A first section will discuss the most recent data concerning the nature of lipomas, their anatomy, clinical presentation, incidence and genetics, which all are necessary for a good understanding of these malformations. A second section will deal with surgery and surgical indications. A third section will develop new hypotheses that could explain the development of lipomas in the human. These hypotheses stem directly from recent studies on avian tail bud growth mechanisms using the quail-chick chimera system [41].

General considerations

The lipoma: nature and behavior

Pathology

In our series, 23% of the lipomas were made up of normal mature adipocytes separated into clusters by numerous collagen bands (Fig. 1). These findings are similar to those already described by many authors [55, 176, 178, 180], who frequently termed these lesions "fibrolipomas" [55, 56]. In 77% of cases the lesion was more complex, including various other tissues originating from ectoderm, mesoderm, or endoderm (Table 1). The most frequent were of mesodermal origin: striated muscular fibers (37%), nerves (63%), and Vater-Paccini and Krause corpuscles (21%). These formations were always intralipomatous and surrounded by dense fibrosis. Tissues of neurectodermal origin were also common, such as glial or neuroglial tissue (18%), arachnoid (7%), and ependymal or glio-ependymal structures (5%). Other tissues were rarer: dermoid or epidermoid cysts, cartilaginous (6%) or bony (3%) nodules, lymphatic (3%) or nervous (1%) ganglia, angiomatous formations (4%), smooth muscle fibers (6%), cavities lined with respiratory or intestinal epithelium (2%), renal glomerules (<1%), and cerebellar tissue (<1%; Fig. 2). In the literature, similar findings were also reported, but rarely [1, 18, 27, 67, 112, 162, 176, 180, 191]. In two cases, the lipoma included endometrial tissue [132, 156].

These findings show that lipomas may be more or less complex lesions and could be considered as either simple or complex teratomas. In the literature, complex forms have been reported less frequently (10–25%) than they have been observed in our experience, perhaps because other series were smaller than ours or samples of the lipomas were insufficient for correct histopathological study.

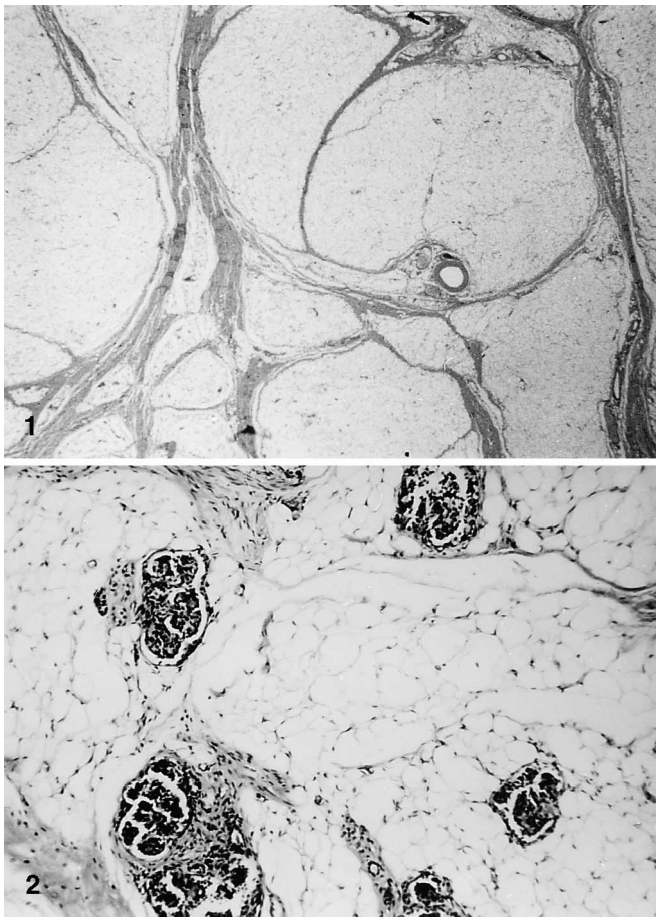


Fig. 1 Photomicrograph of a lipoma. The mass is composed of well-differentiated adipose tissue. Fibrous septa divide the fatty mass. (Haematoxylin-eosin, ×100)

Fig. 2 Photomicrograph of a lipoma showing numerous renal corpuscles. Haematoxylin-eosin, ×200)

Table 1 List of ectopic tissues found within intraspinal lipomas: Necker Enfants Malades (NEM) series

Entoderm	Mesoderm	Ectoderm
Epithelium	Vessel	Nerve
Intestine	Striated muscle	Neuroglia
Respiratory epithelium	Neurosensorial corpuscle	Meninges
Endometrium	Meninges	Ependyma
	Lymphatic ganglia	Cerebellum
	Cartilage	Epidermoid cyst
	Kidney	

Biology

Adipose tissue plays two major roles: lipid storage and lipid mobilization in response to various hormonal and metabolic stimuli. Increased lipid storage and reduced lipid mobilization induce adipocyte enlargement and, in consequence, promotion of fat.

Lipid storage (or lipogenesis) is controlled by lipoprotein lipase (LPL), an important enzyme in lipid and lipoprotein metabolism. LPL is synthesized, glycosylated, and secreted by adipocytes and subsequently anchored to vascular endothelial cells, where this enzyme hydrolyzes the trigliceride core of circulating chylomicrons and very-low-density lipoprotein (VLDL) into free fatty acids (FFA). These FFA are then taken up by the adipocytes to be reesterified into triglycerides. Insulin, the main lipogenic hormone, plays a pivotal role in LPL regulation, because insulin stimulates LPL synthesis, secretion and activity, and also promotes glucose uptake, which is required to allow FFA reesterification inside the fat cell. Lipid mobilization (or lipolysis) proceeds through a complex cyclic AMP-dependent metabolic cascade involving hormone binding to specific receptors located in the cell membrane, adenylate cyclase modulation, intracellular cyclic AMP variation, hormone-sensitive lipase activation and, finally, FFA and glycerol release into the blood. In humans, cathecholamines are the major hormonal signals regulating lipolysis in fat cells. These cathecholamines bind to two different adrenergic receptors displaying opposite effects on lipolysis: the beta-receptor subtype, which stimulates adenylate cyclase, cyclic AMP synthesis and lipolysis, and the alpha2-receptor subtype, which when activated, inhibits adenylate cyclase, cyclic AMP synthesis and lipolysis.

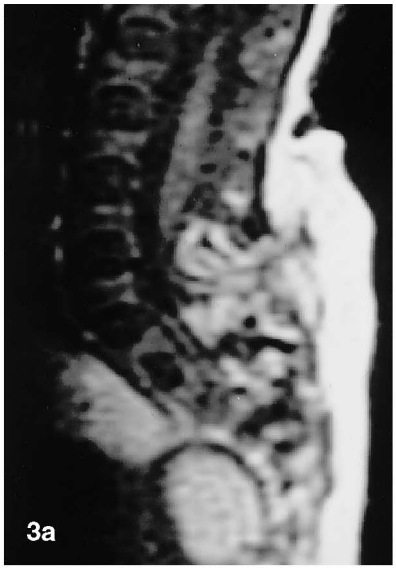
To determine whether the fat cells from congenital intraspinal lipomas behave in the same way as benign subcutaneous lipomas or as normal adipocytes, we have explored the main factors known to be involved in both lipogenesis (LPL and insulin-stimulated glucose incorporation into triglycerides) and lipolysis (numbers of beta- and alpha-adrenergic receptors, adenylate cyclase response to beta- and alpha-adrenergic agonists, cyclic AMP production and lipolytic activities) in the adipocytes from intraspinal lipoma and normal adipose tissue. These studies showed that metabolic activities in the intraspinal lipoma cells were similar to those observed in the adjacent normal adipose tissue [62]. Histologically, congenital intraspinal lipomas also appear to be composed of normal adipose cells. Normal and mature adipocytes have no potential evolution of their own, but are capable of growth as well as regression, along with increase and decrease of the rest of the fatty pool. Their behavior is comparable to that of the normal epidural fat. Epidural fat has also been shown to be capable of growth and of spinal cord compression, not only in Cushing disease but also after high doses of corticosteroids, both of these situations leading to a general increase in adipose tissue. These studies indicate that congenital intraspinal lipomas are radically different from the various known lipomatoses. The latter are, in fact, benign, slow-growing fat tumors, with a proven increase in lipogenesis and defective lipolytic activity.

Growth

It is widely accepted that congenital intraspinal lipomas are anatomically stable lesions. Nevertheless, growth has been reported in 15 cases [8, 9, 13, 16, 18, 73, 92, 118, 119, 130, 150], either subcutaneously or intraspinally [8, 16]. In 14 cases, the increase in size of the lipoma was observed preoperatively, and in 1, postoperatively [72]. In our series, lipomas have been seen to grow in 36 (13.2%) of the patients (Fig. 3), subcutaneously in 28 cases (9%), and intraspinally in 5 others (1.6%). In 3 of these 36 patients (1%), the lipoma recurred postoperatively (Fig. 4), as it did in the case reported by Hall [73]. Among our 36 patients, 21 were infants, 2 were obese adolescents, and 10 were pregnant women. The last 3 had complex teratomas. In all of these cases, lipomatous growth was easily understandable. When obesity occurs, there is a general increase of the fatty pool, including that in the lipoma, which is made up of normal adipocytes. The same process takes place in young infants, in whom adipocytes increase in size [102], to such a degree that the proportion of fat increases from 14% at birth to 25% at 6 months [158]. Lipogenesis also increases at the beginning of gestation owing to secretion of progesterone and estradiol, which accelerate the multiplication of preadipocytes and their differentiation into adipocytes [159]. During pregnancy, the increase in intracellular fluids and variations in venous pressure may also play a part [150]. Rarely, growth of complex teratomas may result from other mechanisms, such as bleeding of intralipomatous endometrial implants [132], malignant degeneration [118, 130], or recurrence of a previously incompletely removed dermoid cyst, as seen in 1 of our patients [149].

The malformation complex

Lumbosacral lipomas have been extensively described [42–44, 149]. They form a heterogeneous group of lesions which have been variously classified [42, 47, 56, 71, 79, 125, 149, 152, 155, 157, 165, 166, 195]. Two anatomical entities should be distinguished: lipomas of the filum, and lipomas of the conus. In our series (Table 2), and also in the literature (Table 3), lipomas of the conus were much more frequent than lipomas of the filum. They also appeared to be more complex. For this reason, these two types of lesion have been studied separately. However, both of them are only part of a wider malformation complex involving the nervous system, the spine, the skin, and often other viscera. The description of these lipomas therefore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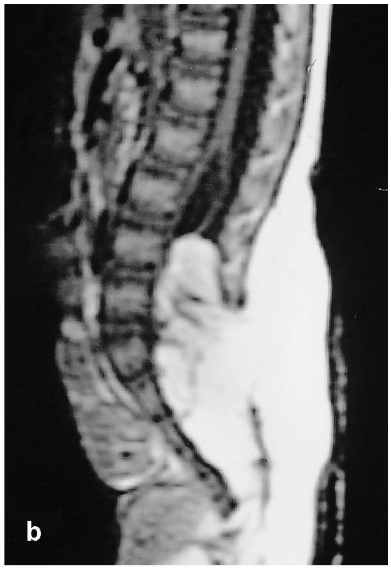
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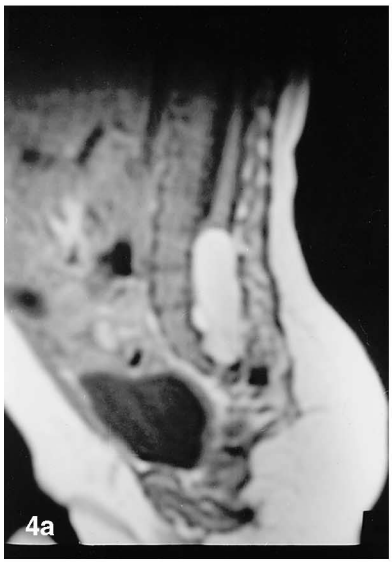
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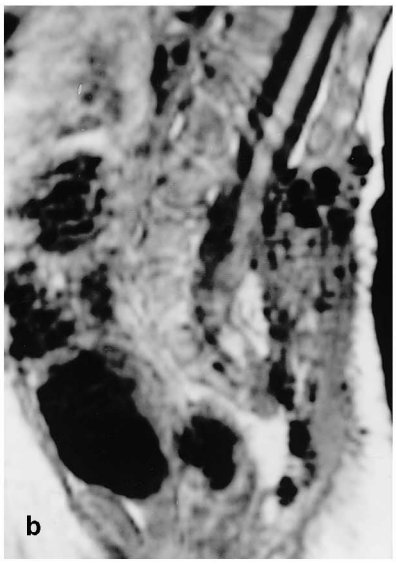
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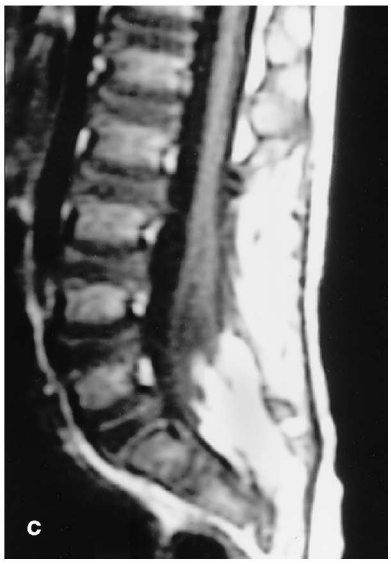
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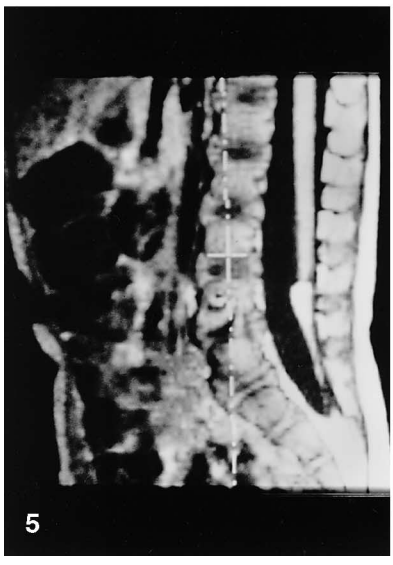
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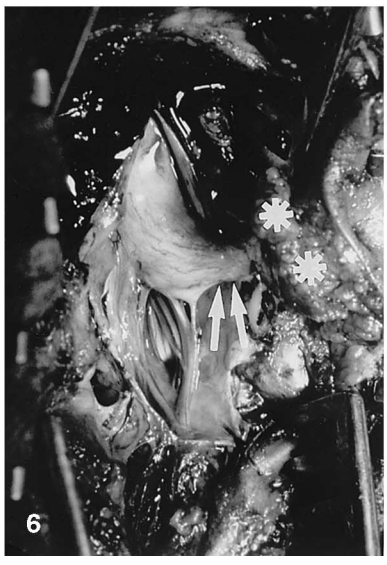
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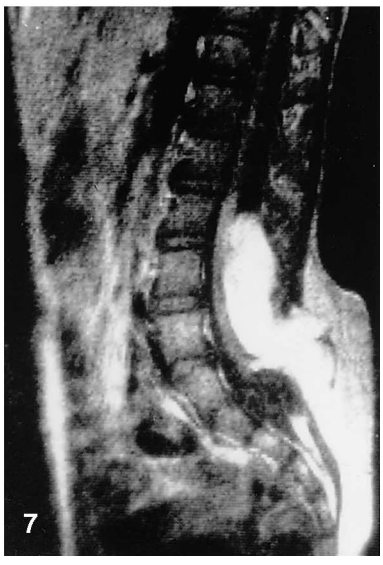
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necessarily requires description of the associated malformations.

The lipoma

Lipomas of the filum. In our series, there were 39 lipomas of the filum. In all but 2 cases (94.8%), the lipoma was intradural, strictly subarachnoid infiltrating a normally localized filum (Fig. 5). In 2 cases, the lipoma, along with the filum, had an abnormal course, spreading out from the dura to extend subcutaneously (Fig. 6). The filum was abnormally thick and rigid in all cases. The spread of the fatty infiltration was variable, involving the whole filum or only part of it.

At surgery, in all but 1 case, the lipomatous filum was surrounded by the apparently normal roots of the cauda equina. In 1 patient, as in the case reported by Brophy et al. [30], some of the sacral roots were intermingled within the malformed filum.

Lipomas of the conus. There were 264 lipomas of the conus in our series. As shown in Table 4, there were numerous anatomical forms, but all had in common the insertion of the lipoma on the terminal cord.

• Typical forms: Typical lipomas of the conus have a tendency to spread out from the dura, extending extraspinally through the defect of spina bifida and finally developing subcutaneously (Fig. 7). In our series (Table 4), these forms were actually in the minority ($n = 98$, 37.1%). The stalk joining the intra- and extraspinal portions of the lesion was frequently adhering strongly to the musculo-aponeurotic edges of the spina bifida. In some patients, this fibro-lipomatous stalk was accompanied by bundles of paraspinal muscles in addition, which also ended on the conus. Intraoperative application of electrical stimuli to these bundles often led to impressive contractions of the cord itself.

The zone of insertion of the lipoma on the cord, or interface, was usually wide. Its upper limit was around L5 in 50% of our patients. Its lower limit was in the sacral region in 90% of them, most often at S3. Its median extension was four vertebral levels.

The volume of intradural lipomas was variable. When they were large, the cord was compressed and the interface pushed away from the dural dehiscence (Fig. 8). When they were small, the cord was suspended from the posterior aspect of the dural sheet and the interface was at the level of

Table 2 Incidence of lipomas of the filum and of the conus: NEM series

	n	%
Lipomas of the filum	39	12.9
Lipomas of the conus	231	76.2
Lipomas of the conus + filum	33	10.9
Total	303	100

Table 3 Lipomas of the filum and of the conus: respective incidences recorded in the literature

Reference	*n*	Conus		Filum	
		n	%	*n*	%
[7]	39	21	54	18	46
[12]	16	12	75	4	25
[32]	42	36	86	3	7
[73]	22	17	77	5	6
[154]	18	14	78	1	5.5
[175]	26	21	81	5	19
Total	163	121	75.1	36	17.1

Table 4 Anatomical forms of lipomas of the conus: NEM series

	n	%
Typical	98	37.1
Subdural, extended to the subcutaneous layers	98	37.1
Atypical	166	62.9
Lipomyelocele	67	25.4
Lipomeningomyelocele	52	19.7
Lipomyelocystocele	6	2.3
Subdural	16	6.1
Subdural associated with a subcutaneous lipoma	4	1.5
Subdural with presacral extension	20	7.6
Total	264	100

Fig. 3 MRI, T1 relaxation time, sagittal views, demonstrating the increase in size of a lumbosacral lipoma between 6 (**a**) and 18 (**b**) months of age. On the second MRI a terminal syringomyelia has appeared

Fig. 4a–c Postoperative „recurrence" of a lumbosacral lipoma. **a** Preoperative MRI demonstrating the presence of a large lipoma extending from L3–L4 to S2. **b** Postoperative MRI 2 months after surgery. **c** Postoperative MRI 18 months after surgery. There is an indisputable increase in size of the residual lipoma in the interval between the last two MRIs. Note the development of a terminal syringomyelia

Fig. 5 MRI (sagittal view) showing a typical lipoma of the filum. The termination of the spinal cord is at L4

Fig. 6 Operative view of a lipoma of the filum (*arrows*) continuing extraspinally into a subcutaneous lipoma (*asterisks*). The superficial orifice of a fistula is visible at the level of the skin

Fig. 7 Typical aspect of a lumbosacral lipoma on an MRI sagittal view: subcutaneous mass, stalk penetrating the spina bifida, intraspinal component of the mass terminating on the conus terminalis. The spinal cord ends at S1

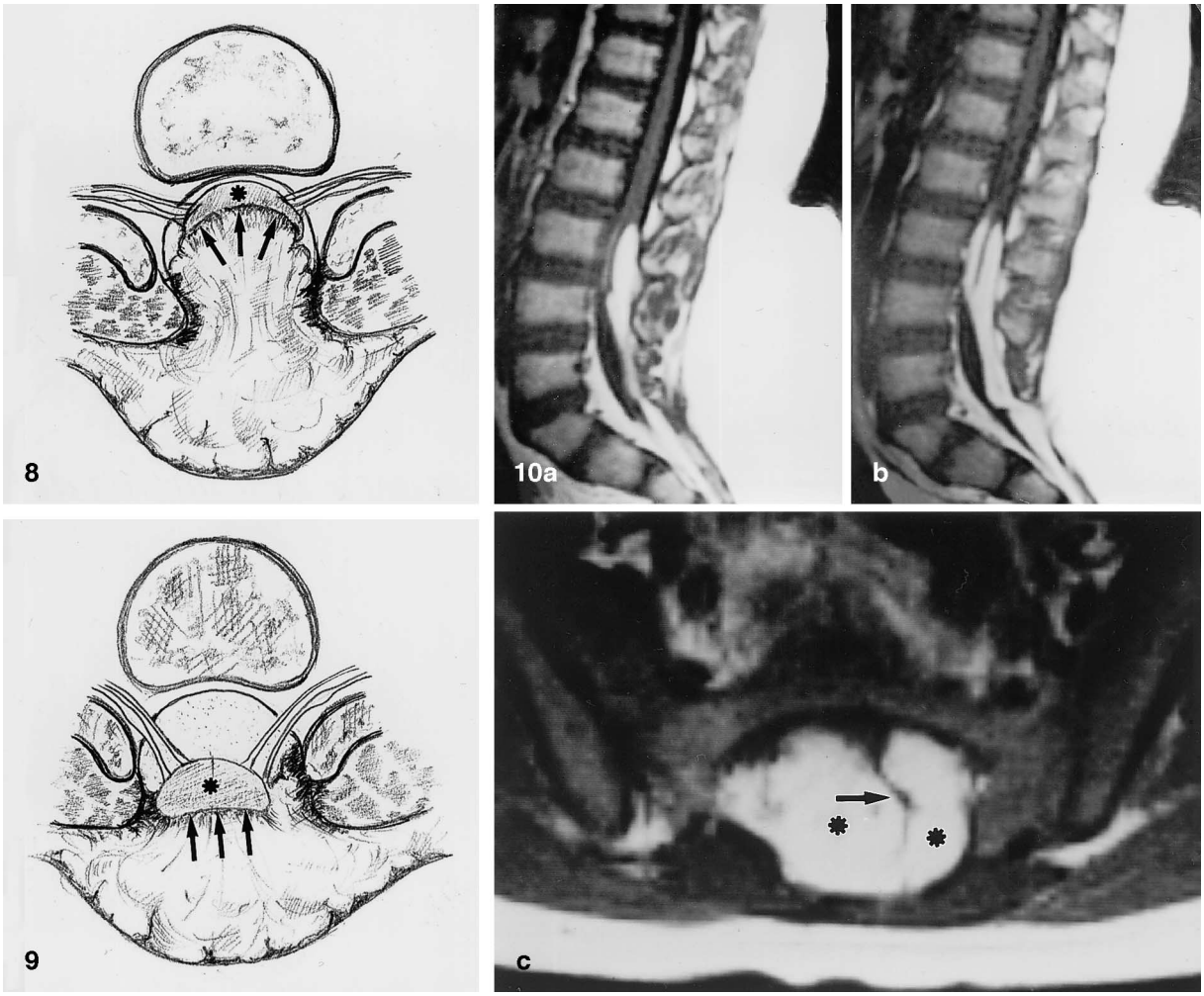


Fig. 8 Schematic drawing showing on an axial view the relationships between spinal cord (*asterisk*), lipoma and dural sac when the cord/lipoma interface (*arrows*) is distant from the dural defect. In such a case the intradural volume of the lipoma is voluminous and attachment of the cord to the dura is loose

Fig. 9 Schematic drawing (axial view) showing the relationships between spinal cord (*asterisk*), lipoma and dural sac when the cord/lipoma interface (*arrows*) is at the level of the dural defect (*arrows*). In such a case the volume of the intradural component of the lipoma is reduced to a minimum, and the spinal cord is firmly attached to the edges of the dural defect

Fig. 10a–c MRI, T1 relaxation time. **a, b** Sagittal median and paramedian views; **c** axial view, showing the invagination of the terminal spinal cord (*arrow*) within the lipomatous mass (*asterisks*)

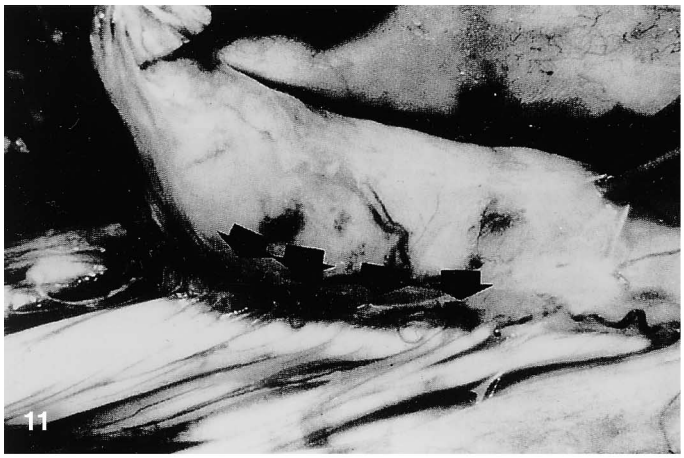
the dural defect (Fig. 9). In these case, adhesions between cord, lipoma and dura were pronounced. Once the lipoma was removed, the interface was usually recognizable by its stronger resistance and whitish color. There is only one description of its histological structure [115]. In this particular case, it was made of connective tissue reinforced by smooth muscles.

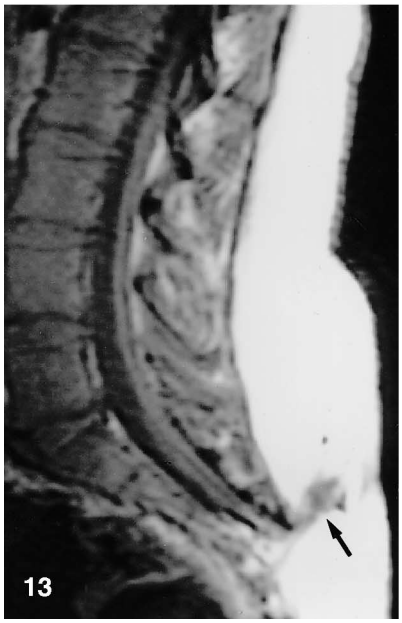
In the majority of cases, the interface was plane. In others, however, it was convex, as shown in Fig. 10, indicating herniation of the dorsal aspect of the cord within the superficial fatty mass.

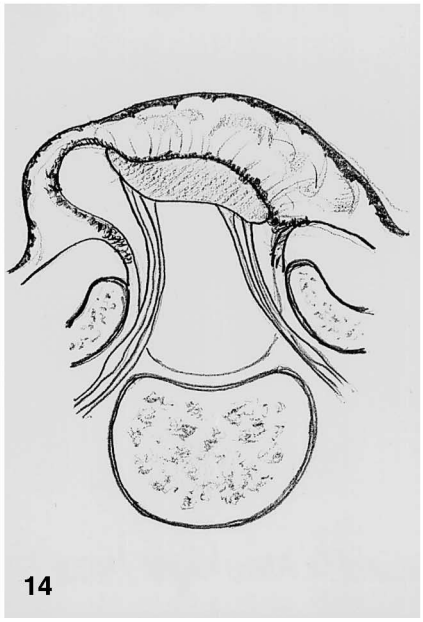
Chapman [42] classified lipomas into four types according to the interface localization on the cord: dorsal, dorso-lateral or lateral, caudal, and dorso-caudal. In our experience, 34.5% of the lipomas of the conus were dorsal, 30% dorso-lateral or lateral, and 35.5% caudal or dorso-caudal. In the dorsal and caudal forms, all roots had an extralipomatous subdural trajectory (Fig. 11). In dorso-lateral forms, in contrast, roots were intermingled within the lipoma and difficult or even impossible to recognize (Fig. 12). The interface, when lateral or dorso-lateral, was located more often on the left side of the cord than on the right ($P < 0.002$).

• Atypical forms: In our series (Table 4), the so-called atypical forms were, surprisingly, in the majority ($n = 62.9\%$).

In *lipomyeloceles*, the cord herniated extraspinally, to end finally within the subcutaneous mass of the lipoma (Fig. 13).







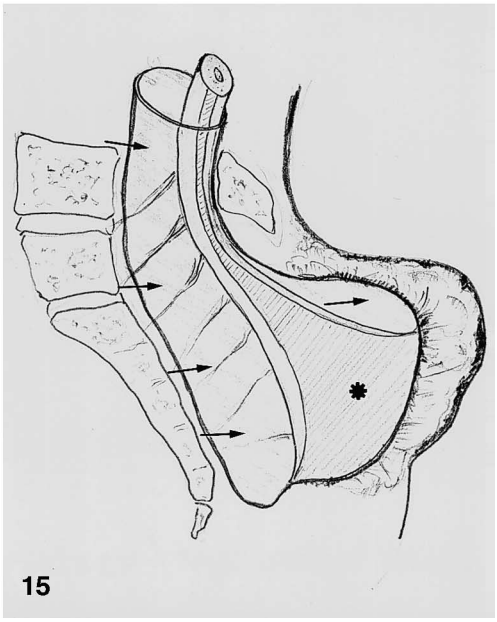


Fig. 11 Operative view showing the insertion of a lipoma on the posterior aspect of the conus terminalis. In this case roots have a normal orientation. All are extralipomatous. The interface between cord and lipoma (*arrows*) is located a few millimetres posterior to the emergence of the dorsal roots

Fig. 12 Operative view showing the posterolateral insertion of a lipoma on the cord. Some of the roots emerge directly from the lipoma (*arrows*)

Fig. 13 Lipomyelocele. On the MRI, the spinal cord extends posteriorly (*arrow*) into the subcutaneous component of the lipoma

Fig. 14 Schematic drawing of a lipomyelomeningocele: both the terminal spinal cord and the dural sac herniate posteriorly. The subcutaneous lipoma is inserted on to the surface of the placode. At this point, there is usually no interposed dura between cord and lipoma. Roots originate from the ventral aspect of the cord and have an exclusively extralipomatous route

Fig. 15 Schematic drawing of a lipomyelocystocele. The terminal spinal cord is distended by a terminal cyst-like syringomyelia (*asterisk*). The inferior end of the cavity is occluded by the subcutaneous lipoma and the dura. The syringomyelia does not communicate with the subarachnoid spaces (*arrows*)

Lipomyelomeningoceles were characterized by the association of a subcutaneous meningocele to the lipoma (Fig. 14).

Lipomyelocystoceles (Fig. 15) were much more rare (2.3%). In these cases, the terminal cord was enlarged by the presence of a pseudo-cystic terminal hydromyelia closed superficially by the dura of the cul de sac, itself covered by the lipoma. As already described [125, 188] the intracystic and subarachnoid CSFs never communicated freely. The pseudocyst membranes were described as having an ependymal structure [49, 125, 143, 188]. In our series, postnatal MRI consistently failed to yield the correct diagnosis of the malformation. In 1 case, the diagnosis was established prenatally on fetal ultrasonography.

Strictly subdural lipomas were more rare in our series (6.1%) than in the literature, where they account on average for between 7% [32] and 19% [175]. As in other series, they were usually not associated with spina bifida.

Finally, 6.6% of the lipomas had a presacral extension (Fig. 16), usually being in close contact with the rectum. In some cases, the cord also herniated presacrally.

Associated malformations

Associated malformations were very frequent, involving the cord and roots, the spine, the skin, the urogenital and anal apparatus, and other viscera to varying degrees.

Intraspinal malformations

• *Cord and root anomalies:* The cord and root anomalies found in our patients are displayed in Table 5. Except for hydromyelia, they were significantly more frequent in lipomas of the conus than in lipomas of the filum. Root disposition (horizontal or vertical) was not taken into account, neither being a true malformation, but rather the consequence of the low-lying conus. The conus was abnormally low in 94.8% of our cases. It was in a normal position in 5.2% of them, with a similar incidence for lipomas of the conus and of the filum. This percentage of normally ending cord in the case of lipomas of the filum was higher in the series of Warder and Oakes [192] and in that of Khoury et al. [100], respectively 28% and 87%.

Root anomalies were often multiple in the same patient. They were noted in 168 of the 291 patients operated on (57.7%); all but 1 had lipomas of the conus. In the literature also, lipomas of the filum are rarely associated with root malformations. There is only 1 reported case [30] in which sacral roots, as in 1 of our patients, were intermingled within the lipomatous filum. As already reported [16, 128, 163], intralipomatous and short roots often made surgery difficult. In the first situation, roots could hardly be differentiated from intralipomatous fibrotic bands (Fig. 17). In the second, they always prevented the surgeon from correctly freeing the cord on the side of the anomaly (Fig. 18).

An intraspinal cavitation existed in 61 of our 303 patients (20.1%) with similar incidences for lipomas of the filum (8 of 39; 20.5%) and lipomas of the conus (53 of 264; 20%). In the literature, this incidence ranged from 11% [70] to 27% [83]. As in many other series [8, 68, 70, 79, 83, 93, 103, 115, 166], syringomyelia was most often localized at the terminal cord. In our patients it extended on average over two vertebral levels. Dorsal, cervical and bulbar hydromyelia in our series were noted, respectively, in 3.3%, 0.6%, and 0.3% of cases.

Diastematomyelia or diplomyelia were observed in 24 of our 265 lipomas of the conus (9%), and in 1 lipoma of the filum. This incidence is lower than those reported by Stolke [175] and Lassman and James [111], which were respectively 15.3% and 26.9%. In 1 of our cases, the cord had a 180° twist, so that on preoperative MRI the lipoma was thought to be anterior to the cord.

• *Multiple intradural lipomas:* Some lipomas of the conus reported in the literature were associated with lipomas of the filum [168] or with lipomas of the cord at a higher level, either unique [66] or multiple [186]. In our series, a lipoma of the conus was associated with a lipoma of the filum in 33 patients (10.9%), and with a dorsal lipoma of the cord in one.

• *Intralipomatous mass:* An intralipomatous mass was reported in 20 of our patients (9%), most often in the case of lipoma of the conus. There were 3 dermoid cysts, 9 dermal sinuses, 7 angiomas, and 1 teratoma (Fig. 19). In 1 patient, the intralipomatous angioma extended on to the cord and was associated with a large metameric cutaneous angioma.

• *Dural and subdural malformations:* Three patients had a dorsal arachnoid cyst. In the literature, associated intradural malformations were rarely reported: 1 enteric cyst [166], 2 dural arteriovenous malformations [52, 156], and 1 intramedullary mature teratoma [144].

Spinal malformations. In our series the spinal malformations were dominated by spina bifida, as they were in the literature (Table 6). Its frequency was slightly higher in lip-

Table 5 Cord and root malformations in patients with lipomas of the filum and of the conus: NEM series (291 patients treated surgically)

Malformations	Filum ($n = 38$)		Conus ($n = 253$)		*P*
	n	%	*n*	%	
Cord					
Low-lying					0.014
L3-5	22	58	90	36	
S1-5	14	37	148	58	
Duplicated	0	0	24	9	0.000
Hydromyelic	8	21	53	21	NS
Rotated	0	0	1		
Roots					0.0001
Intralipomatous	1	3	100	40	
Infiltrated	0	0	25	10	
Agenetic	0	0	35	14	
Dysgenetic	2	5	78	31	
Short	0	0	67	26	
Total (no. of patients)	1	2.6	167	66	

Table 6 Incidence of spinal malformations

Series	Spina bifida		Sacral agenesis		Spinal deformities	
	No. of patients	%	No. of patients	%	No. of patients	%
NEM	303	91	303	20	303	14.6
Literature	148	69	85	27	224	9.5
Total	453	80	388	23.5	527	12

Fig. 16 Lipoma of the conus terminalis associated with sacral agenesis. The MRI, on a sagittal view, shows that the lipoma extends anteriorly and is in contact with the rectal ampulla

Fig. 17 Operative view during resection of a lipoma. The intraoperative stimulation of an intralipomatous tractus (*arrow*) gave responses in S1 territory on one side

Fig. 18 Operative view after the resection of a lipoma of the conus. The terminal cord remains attached to the left aspect of the dural sac owing to the presence on the same side of abnormally short roots. Roots on the right side are still under tension

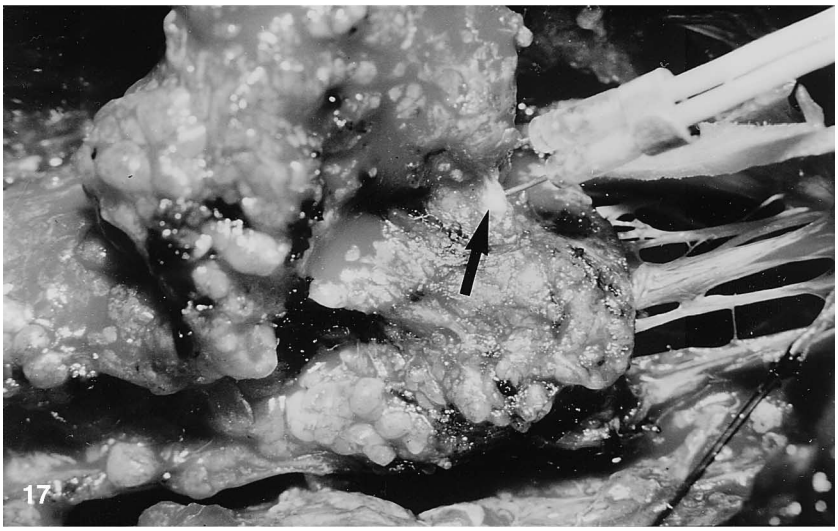
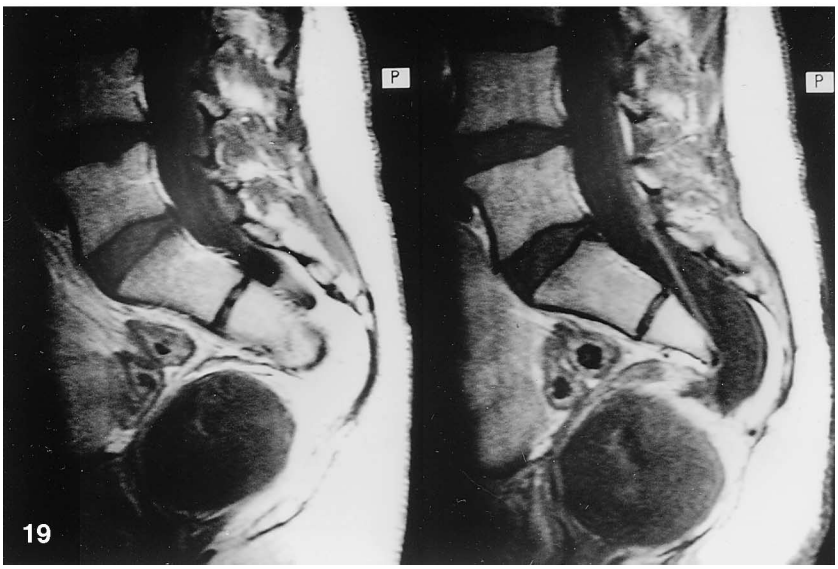


Fig. 19 MRI, sagittal views, T1 relaxation time showing a teratoma included within the presacral extension of a lipoma



omas of the conus (254/264; 96.2%) than in lipomas of the filum (32/39; 82%), without, however, attaining statistical significance. In 80% of our patients, its upper level was L5 or S1. In 90% of them, the whole sacrum was involved, a similar proportion to that in the series of Naidich et al. [136].

The frequency of sacral agenesis is approximately 25% of all cases. As reported by others [25, 46, 48, 51, 79, 96, 107, 136, 177, 193], however, its incidence was higher in patients with uro-genito-anal malformations (93%).

Scoliosis was related to malformations of the vertebral bodies or shortening of one lower limb in all but 1 of our patients. This leads us to think that in lipomas this is probably not due to spinal cord tethering as in meningomyeloceles [128].

Other spinal anomalies were rare: diastematomyelia (8%), hemivertebrae (7%), vertebral fusion (6%). Theandeer reported one case with malformations of the iliac bone and homolateral skeleton [183].

Brain malformations. Data from the literature are difficult to assess and compare, because most groups include all types of occult dysraphisms in the same study. However, in all series, brain malformations are very rarely associated with lipomas, which is strikingly different from what is commonly observed with meningomyeloceles. Their incidence in our series was 3.6% (11/303). There were 4 Chiari malformations (1.3%), 4 cases of hydrocephalus (1.3%), 2 of which were due to aqueduct stenosis, 2 hemispheric arachnoid cysts (0.6%), and 1 Dandy-Walker malformation. Of the 4 Chiari malformations, 1 was particularly interesting, in that the malformation was absent at birth and developed with time to become symptomatic at the age of 20 years [149]. In the literature, the following malformations have been reported, sometimes in the same patient: encephaloceles [42, 157, 166], Chiari [108, 166], callosal agenesis [189], intracranial lipoma [157], aneurysms and arteriovenous fistulas [157].

Extra-axial malformations. Visceral malformations other than of the brain and of the cord were reported in 16% of the published cases [32, 79, 93, 110, 116, 119, 141]. As shown in Table 7, this is less than we have found (27%). Uro-ano-genital malformations were the most frequent. One observation was very rare: the child presented with an extra upper limb [149], a case similar to those reported by Parkinson and Humphreys et al. [80, 142].

Skin anomalies. Cutaneous abnormalities were present in 89% of our patients which is in accordance with all data in the literature (Table 8). They were dominated by the presence of a subcutaneous lump indicating the existence of the underlying lipoma, but were commonly multiple in the same patient.

Table 7 Associated malformations (NEM series)

Malformations	*n*	%
Ano-rectal	20[a]	6.6
Anal imperforation	10	
Anorectal stenosis	7	
Anal malposition	4	
Complex malformations	2	
Genital	12[a]	4
Vulva	7	
Uterus	4	
Cloaca	1	
Penis and testis	1	
Urinary	19[a]	6.3
Unilateral kidney/agenesis	7	
Duplication	6	
Kidney malposition/dysmorphism	4	
Bladder extrophy	1	
Reflux	1	
Cardiac	7[a]	2.3
Atrial septal defect	3	
Ventricular septal defect	1	
Patent ductus arteriosus	1	
Pulmonary stenosis	1	1
Tetralogy of Fallot	1	
Other	27[a]	9
Limb	8	
Rib	5	
Angiomatosis	4	
ENT	3	
Microphthalmia	1	
Skeletal herniatrophy	1	
Recklinghausen	1	
Teratoma	1	
Total	82[a]	27

[a] No. of patients

Table 8 Associated cutaneous abnormalities

Series	*n*	%
NEM study	303	89
Literature	611	92
Total	914	90.5

Genetic disease. A case of Turner syndrome [133] and one of trisomy 21 [79] have been reported in the literature with no mention of the type of lipoma.

Incidence in the population

In the neurosurgical literature, the only reported data concern the respective incidence of lumbosacral lipomas vs meningomyelocele [19, 94, 189] or spina bifida [7, 12, 36, 91, 111]. These data are of little interest, since they reflect only personal experience.

Data obtained during routine autopsies on patients in whom spinal cord disease is not suspected (Table 9) are more important, but the incidence of incidentally found lipomas ranged from 0% (Schlesinger cited by Stookey [176]) to 6% [124], with an average of 0.03%. Interestingly, lipomas of the conus were almost 10 times as common as lipomas of the filum, which contradicts all clinical experience.

Recent studies aiming to determine the prevalence of lumbosacral lipomas retrospectively by MRI in adults in whom disc disease or lumbar stenosis was suspected showed a 1.5–5% incidence of lipomas. All were lipomas of the filum, and the majority were small [31, 50, 131, 139, 187].

We tried to approach the clinical incidence of lipomas by examining two French regional registers [149]. When lipomas of the filum and of the conus were considered together, their incidence ranged from 4/100,000 to 7.8/100,000. These were underestimated, however, since they were calculated from a population of neonates whose lipomas had been clinically diagnosed at birth, that is, from babies born with visible cutaneous stigmata or neurological deficits. It is possible to minimize this underestimation by taking into account the 10% of lipomas in all series that present with no cutaneous stigma. When this is done, the minimal incidence of lipomas in France would range between 4 and 8.5/100,000. This incidence is low, and much lower than that reported at autopsy or radiologically. This discrepancy could be explained by the existence of quite a large number of asymptomatic patients.

Familial data

A family study was performed in 71 patients in our series. In 29 of these 71 cases, lipomas were associated with other anomalies: sacral agenesis in 7, sacral and uro-genital abnormalities in 10, other malformations without sacral agenesis in 12. Plain X-rays of the spine were performed in all parents to search for spina bifida or other spinal abnormalities. Malformations at the lumbosacral-sacral junction, such as lumbalization or sacralization, were not taken into account owing to their high frequency. The aim of this study was to compare the frequency of spina bifida occulta in the relatives of our patients with that in the normal population. Spina bifida occulta was found in 69% of these relatives irrespective of the type of lipoma in our patients. This frequency is higher than that observed in the normal population, which is estimated to range from 25% to 30% [25, 40, 177].

Some familial cases were reported in the literature, indicating the possibility of an autosomal dominant mode of inheritance. These cases included different types of the dysraphic state. Among them, the specific Currarino syndrome must be emphasized, in which lipoma is associated with uro-genital and perineal defects [51]. This syndrome could be linked to others without lipomas. Similar abnormalities were described in mice with *T* locus mutation, and Fellous et al. [57] described a large family with spina bifida occulta and sacral agenesis linked to *HLA* locus. Furthermore, Lynch et al. [120] showed a deletion of chromosome 7q36 in probands with sacral agenesis, while others [121] showed a positive linkage in this region in a large family.

Therefore, it appears that lipomas, occult spina bifida, sacral agenesis and caudal abnormalities could be related to the same genetic factors. However, in our study, lipomas were an apparently sporadic event in 30% of cases.

Preoperative clinical presentation

Data in this section stem from our series (NEM) and the literature (Table 10). Numbers and percentages must be interpreted with caution, because they have been obtained from various and often inhomogeneous series. Nevertheless, they are presented for comparison with our results. Except for data concerning gender, they cannot be interpreted as a metaanalysis.

Sex ratio

In our series, as in most others in the literature (Table 10), there was a significant female preponderance (F/M: 1.6/1;

Table 9 Incidence of incidentally discovered lipomas at autopsy

Reference	No. of autopsies	Lipomas		Filum		Conus	
		n	%	*n*	%	*n*	%
[17]	300	4	1.3	3	1	1	0.3
[113]	90	5	5.5	0	0	5	5.5
[124]	47	3	6	0	0	3	6
[154]	650	1	0.1	0	0	1	0.1
Schlesinger (cited in [176])	35000	0	0	0	0	0	0
Total	36087	13	0.03	3	0.008	10	0.02

Table 10 Series taken from the literature

Reference	Year	Age	No.	Female	Male	Sex ratio (F/M)
[129]	1930	Children	9	7	2	3.50
[18]	1950	Mixed	9	9	0	
[91]	1956	Mixed	8	6	2	3.00
[84]	1962	Mixed	16	10	6	1.67
[178]	1962	Mixed	9	7	2	3.50
[53]	1965	Children	12	8	4	2.00
[63]	1966	Literature	95	44	51	0.86
[111]	1967	Mixed	26			
[36]	1968	Children	90	62	28	2.21
[123]	1972	Mixed	8			
[71]	1973	Mixed	22			
[185]	1973	Mixed	37	19	18	1.06
[155]	1974	Mixed	36	23	13	1.77
[7]	1975	Children	73	49	24	2.04
[189]	1976	Mixed	13	9	4	2.25
[172]	1977	Elderly	3	1	2	0.50
[4]	1979	Children	2	2	0	
[32]	1979	Children	42			
[122]	1979	Mixed	13			
[145]	1979	Mixed	9	4	5	0.80
[61]	1980	Mixed	46	25	21	1.19
[76]	1982	Children	86	49	37	1.32
[117]	1982	Mixed	21			
[141]	1982	Mixed	23	13	10	1.30
[44]	1983	Children	14	10	4	2.50
[153]	1983	Mixed	102	46	56	0.82
[168]	1983	Children	76			
[87]	1984	Children	10	3	7	0.43
[109]	1985	Mixed	61	26	35	0.74
[79]	1985	Children	97	60	37	1.62
[196]	1985	Mixed	55	33	22	1.50
[126]	1986	Children	50	29	21	1.38
[160]	1986	Children	6	4	2	2.00
[71]	1986	Children	38	22	16	1.38
[116]	1987	Mixed	29	19	10	1.90
[95]	1988	Children	20			
[97]	1988	Children	40	23	17	1.35
[175]	1988	Children	26	16	10	1.60
[30]	1989	Children	25	19	6	3.17
[134]	1989	Mixed	20	7	13	0.54
[166]	1989	Mixed	40			
[59]	1990	Children	31	20	11	1.82
[75]	1990	Mixed	25	7	13	0.54
[94]	1990	Mixed	80	51	29	1.76
[100]	1990	Mixed	19	11	8	1.38
[128]	1990	Children	90			
[140]	1990	Children	61			
[186]	1990	Children	47			
[11]	1992	Children	35	23	12	1.92
[2]	1993	Adults	5	1	4	0.25
[77]	1993					
[164]	1993	<1 year	18			
[47]	1994	Children	98			
[83]	1994	Mixed	143	86	57	1.51
Total			2069	863	619	1.60

$P = 0.0001$). Interestingly, this differs from the sex ratio for meningomyeloceles, where there was an equal sex distribution, and for spina bifida occulta, where there is a male preponderance [40, 177].

Age at diagnosis

In our series, the average age at diagnosis was 6.4 years, which is similar to that reported in the literature (7.1 years). As already emphasized by others [32, 116], the malformation was diagnosed earlier in the presence of cutaneous stigma. There has also been a trend to earlier diagnosis in recent years. From 1990 to 1994, the average age at diagnosis was 9 months in the presence of skin abnormality.

Age at onset of deficits

The average age at onset of neurological deficits was 5 years in our patients. In the literature, it ranges from 5 to 10 years. In the NEM series, deficits were present at birth in 22% of cases.

Symptoms

In all series, the two main clinical features leading to diagnosis are cutaneous stigmata on the lower back and neurological disturbances, mainly neuro-orthopedic and sphincter-related.

The cutaneous syndrome

Cutaneous anomalies were present in 89.4% of our patients. When all series (NEM and the literature) are considered as a whole, 90.5% (Table 8) of the patients have skin stigmata, which are not only of various types, but also frequently multiple in a same patient (Fig. 20). As shown in Table 11, the most frequent of these skin anomalies was, in our patients, a subcutaneous lump located at the midline of the lower back. Usually homogeneously soft, indicating the presence of an underlying lipoma, the lump was fluctuating in some cases, owing to the presence of an associated meningocele. When the subcutaneous lipoma was situated low down, the lump was replaced by an intergluteal fold deviation. Cutaneous stigmata were most often on the midline ($n = 219$; 72.2%), but when lateral they were predominantly on the left side (75% of cases; $P < 0.001$). In 3 of our patients, as in some in the series of Lassman and James [111], meningitis resulting from dermal sinus revealed the lipoma. Some cutaneous abnormalities are rare, such as of our cases metameric cutaneous angioma, which was seen in 1 of our cases extending deep into a large lipoma of the conus [149].

Most authors consider that skin anomalies have a considerable diagnostic value [3, 24, 65, 73, 74, 169, 181], and that their absence is generally the reason for delayed diagnosis [116]. This explains why, in the literature as well as in our series, adolescents and adults had minor or atypical cutaneous anomalies or none at all.

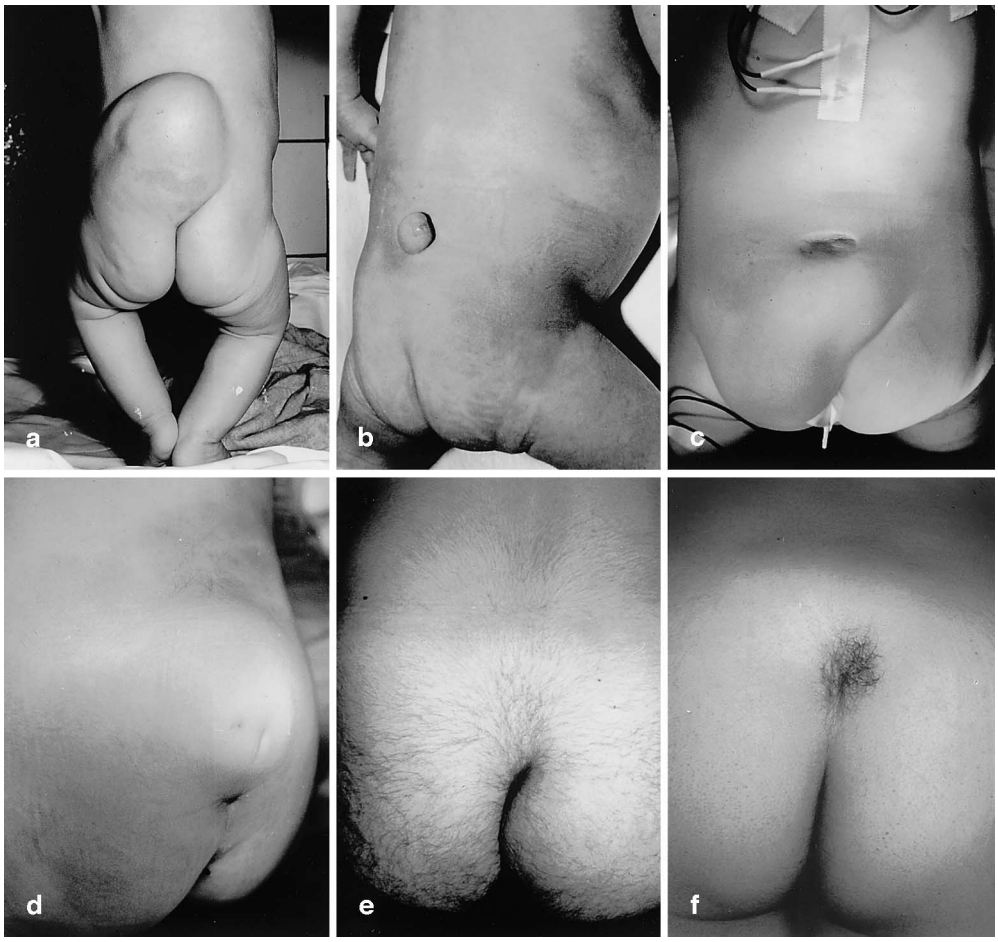


Fig. 20 a–f Some cutaneous stigmata associated with an intraspinal lipoma

Table 11 Types of cutaneous abnormalities: NEM series

Abnormalities	*n*	%	Comments
No anomalies	34	11	
At least 1 anomaly	269	89	221 discovered at birth
Subcutaneous lump	192	64	Including 66 isolated
Angioma	89	29.6	Including 8 isolated
Umbilication	50	16.6	Including 15 without lipoma
Intergluteal fold deviation	50	16.6	Including 17 without lump
Cutaneous aplasia	27	9	Including 15 without lipoma
Dermal sinus	23	7.5	Including 13 without lipoma
Caudal appendix	20	6.6	Including 5 without lump
Patchy hair	18	6	Including 11 without lipoma
Hypertrichosis	11	3.6	100% diastematomyelia

In our patients, coccygeal dimples were frequently associated with subcutaneous lipomas (Fig. 20 d). The current notion that these dimples, when isolated, are never associated with a dysraphic state, was not true in 2 patients, who had associated lipomas of the filum (Fig. 21). The incidence of such an association is unknown, and probably

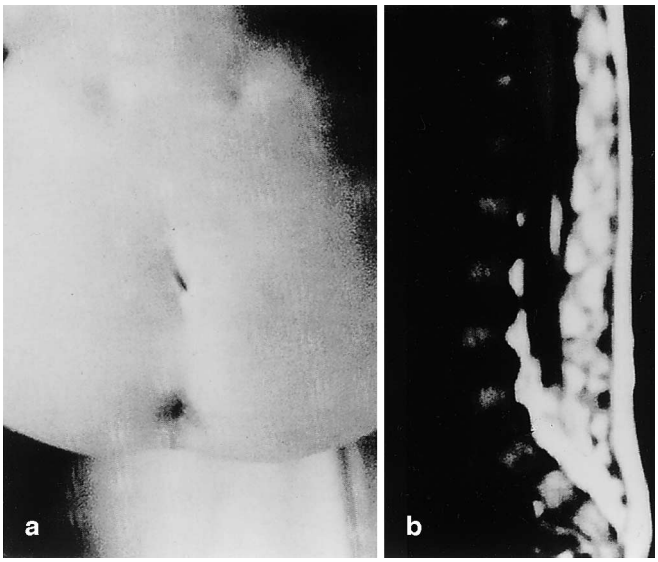


Fig. 21 **a** Typical coccygeal dimple. **b** Same patient, MRI, T1 relaxation time, sagittal view. Presence of a lipomatous filum

it is rare. Routine MRI to investigate isolated coccygeal dimples would be unreasonable, if only for economic reasons, but routine ultrasonography under the age of 6 months would be advisable.

To assess the real diagnostic value of each of these skin abnormalities better, we studied 36 consecutive patients admitted to the dermatology department of Necker-Enfants Malades Hospital for lumbosacral skin anomalies from 1990 to 1994 [149]. An intraspinal lipoma was diagnosed in 5 of these 36 children (13.8%). In 4 of the 5, skin abnormalities were multiple, but they were dominated by a subcutaneous lump in 3, a caudal appendage in 1, and a deviation of the intergluteal fold in 1. Among the 31 children without intraspinal lipoma, subcutaneous abnormalities were usually isolated. Interestingly, this was the case for the 14 with isolated cutaneous angioma. The conclusions that we drew from this study were that (1) cutaneous abnormalities of dysraphic nature are usually multiple, (2) isolated angiomas are not indicative of an underlying dysraphy, and (3) the significance of the intergluteal fold deviation when isolated is not clear, since only one fourth of the children so affected had an intraspinal lipoma.

The neurological syndrome

Problems. The extreme difficulty of documenting neurological normality in neonates or infants and neurological deterioration in adolescents must be emphasized.

In the very young, sensory impairment, motor deficits when discrete and localized in the plantar sole muscles, voiding difficulties in the absence of definite incontinence or of repeated urinary infections may remain clinically undiagnosed. Pain, for example, which was reported in 33% of the adults [141], was noted in only 13.3% of our children, and never reported by others in the literature.

In young children and adolescents, neurological deterioration is often difficult to recognize or to confirm. From 3 to 6 years of age, persisting walking instability, persisting incontinence, or development of a club foot may indicate a recent aggravation, but may also reveal a previously undiagnosed deficit. Similarly, in adolescence, the progressive deformation of a known club foot or the development of trophic ulcerations does not necessarily signify a progressive neurological aggravation. The former can result from the mere skeletal growth of the foot, and the latter from old, stable but previously undetected sensory troubles. All these difficulties explain why, in our patients, an EMG and a urodynamic study were routinely performed not only preoperatively, but also twice after surgery, once soon after, and the second at the age of 3–5. Furthermore, these investigations were subsequently repeated if the development of neurological deficits was in question.

In all series together, neurological deficits affect more than 58% of the patients (Table 12) and are dominated by sphincter disturbances (Table 13).

In our series, as well as in those of others [74, 78, 93], the number of asymptomatic patients decreased with age (Fig. 22). This was interpreted as indicating that the older the patients, the worse their neurological condition. In our opinion, such a conclusion cannot be drawn until the number of asymptomatic adults is known; they of course are not seen in the hospital and therefore do not appear in medical statistics. Our conviction is that, at present, curves such as in Fig. 22 only indicate that adults who become symptomatic or deteriorate require care and hospitalization.

Sphincter disturbances. Overall in all series, vesical sphincter dysfunctions are the most frequent deficit (Table 13). They affected more than half of our patients, and this percentage was even higher in the case of sacral agenesis (69%) and in adulthood (80%).

Micturition difficulties were more frequent than any other type of sphincter disorder (Table 14). In contrast to the situation with meningomyeloceles, incontinence was rarely passive resulting from a flaccid bladder and sphincter paralysis, but mainly the consequence of dysuria, pollakiuria, urgent miction, and incomplete voiding. Bladder infections secondary to urinary retention were also frequently observed.

Preoperative urodynamic studies were performed in 112 patients. They were normal in 48 (42.8%) and abnormal in 64 (57.2%). When abnormal, they revealed a hyperactive bladder in 50 cases (44.6%), a hypoactive bladder in 7 cases (6.2%), and a mixed bladder in the remaining 7 (6.2%). In

Table 12 Incidence of neurological abnormalities

Series	No.	%
NEM study	303	56.7
Literature	661	61
Total	964	58.8

Table 13 Preoperative neurological symptoms

Series	No. of cases	Motor		Sphincteral		Sensory		Pain	
		n	%	*n*	%	*n*	%	*n*	%
NEM	303	95	31	127	42	84	27.7	40	13.2
Literature	441		39		62				
Total	744		35		52		27.7		13.2

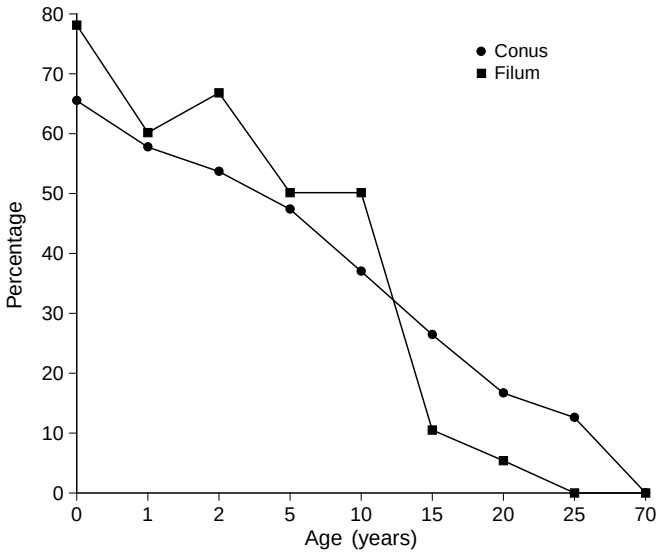


Fig. 22 Percentage of asymptomatic patients by age

Table 14 Types of uro-anal dysfunction: NEM series

Types of problems	*n*	%
Urological		
Incontinence	61	20.1
Dysuria/retention	59	19.5
Infection	52	17.2
Renal insufficiency	5	1.7
Fecal		
Incontinence	35	11.6
Retention	37	12.2
Anal tonus		
Hypertonic	8	2.6
Hypotonic	77	25.4

78% of the patients, the hyperactive bladder was associated with a vesico-sphincteral dyssynergia.

All cases with hypoactive or mixed bladder were clinically abnormal. In contrast, 18 of the 50 patients with a hyperactive bladder were asymptomatic (36%). Among these 18 cases, 15 were children under 2 years of age. Similar findings have been reported in the literature [14, 61, 69, 100, 164]. They illustrate the difficulty of both correctly detecting sphincter disturbances and interpreting urodynamic studies in the very young, in whom both incontinence and bladder hyperactivity are physiological [97, 164].

Bowel dysfunction is rarely reported in the literature [39, 185]. In our patients, constipation was more frequent than diarrhea and sometimes so severe that digital evacuation of the rectum was necessary. Stool leakage was more often due to stercoral retention than to sphincter hypotonia, and all patients with such problems also had urinary disorders.

Sexual dysfunction. Although rarely reported, sexual disorders are probably not rare in adults. They were present in 25% of Thomas and Miller's patients [185] and were reported by many of the adults in our series. As already noted by Thomas and Miller [185], women mainly complained of insensitivity during sexual intercourse, and men of weak erections, sometimes associated with urinary leaks during ejaculation. No case of sterility was reported in the literature.

Neuro-orthopedic syndrome. This syndrome was present in 31% of our patients. This percentage is lower than those reported in the literature (Table 13). As already described by Lassman and James [111], it affected the lower limbs, and to a lesser degree the spine. Paralysis and sensory deficits usually predominated distally: 40% of our patients had a club foot, often with an equinovarus deformity with clawing of the toes (Figs. 23, 24), 36% had muscular atrophia of the leg, 21%, hypotrophia of the leg or foot, and 27.6%, superficial hypoesthesia. Trophic ulcerations secondary to deep anesthesia were much rarer (5.6%). We never observed hypertrophy of the leg as reported by Bertoni et al. [22]. At examination, 34% of the patients had a central motor neuron syndrome. Pain, as described by Pang and Wilberger [141], was diffuse in the whole limb or in a segment of the limb, and affected a radicular pattern in only one of our patients. In the literature, there is only 1 other case mimicking sciatica from a disc hernia [64].

In our experience, these problems were rarely as extensive as in the presence of meningomyeloceles. Their upper level never exceeded L4, and the same observation was made by Kanev et al. [94]. They were often unilateral, or at least asymmetrical, predominantly on the left side (58%; $P < 0.005$). In the case of asymmetrical vertebral abnormalities, neurological problems also predominated on the side of these abnormalities.

Preoperative EMGs were performed in 121 patients. Nearly half of them were under the age of 1 year. Of these patients, 73 (60.4%) were clinically apparently normal, and 48 (39.6%) had deficits. In asymptomatic patients, EMGs were normal in 35 (55.6%), but altered in 28 (44.4%). In no case, however, was denervation severe. In symptomatic patients, signs of denervation were constant and severe, mainly affecting the feet and perineum. For the 76 patients with abnormal EMGs, signs of denervation at the anal sphincter predominated. The highest abnormal level was L2 in 2% of the cases (higher than clinically expected), L4 in 11%, L5 in 22.5%, S1 in 16%, and S2 in 36%. Signs of denervation were symmetrical in 54.5% of the cases, predominantly on the right side in 22.5% and on the left side in 23%. Central motor neuron dysfunction was constant at the anal sphincter and perineum, but found in lower limbs in only 21% of cases.

Currarino syndrome. Kennedy in 1926 [99], Ashcraft and Holder in 1974 [10] and Currarino et al. in 1981 [51] de-

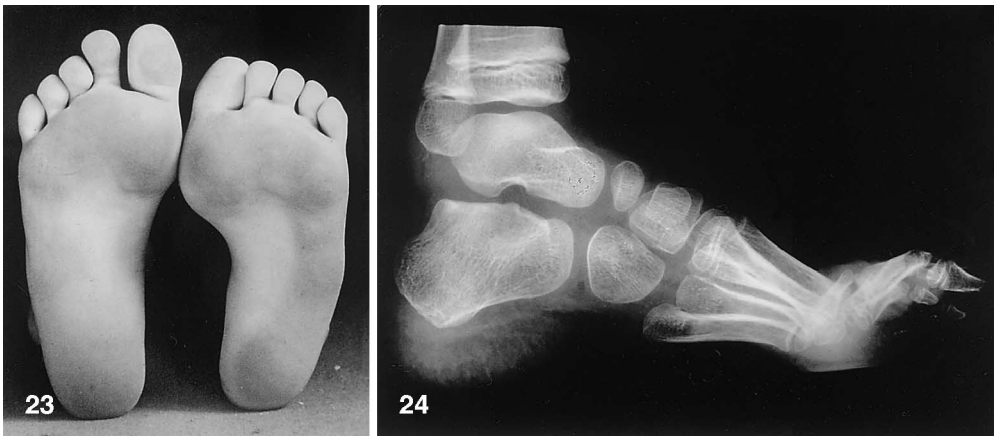


Fig. 23 Typical hypotrophic club foot in a patient with lumbosacral lipoma

Fig. 24 Neuroorthopaedic syndrome: X-ray of the foot showing clawing of the toes

scribed a syndrome associating sacral agenesis, presacral mass and perineal malformations. The presacral mass can be a meningocele, a teratoma, an enteric cyst or a lipoma [51, 107]. This syndrome presents some analogies with that of caudal regression [54], which is associated with various degrees of malformations of the lumbosacral spine, of the lower limbs, and of the uro-genito-anal tract. Moreover, it raises genetic and embryological problems. It is rare, so that is has generally been reported in case reports. In large series of lipomas its incidence varies from 1.8% to 5.1%. In ours it was 5.3% (16 of 303 cases). The diagnosis of the syndrome is usually made at birth or soon after, owing to the perineal malformations. This was the case in 75% of our patients, in 53% of whom the revealing symptom was constipation.

Cutaneous anomalies were present in 60% of the cases in our series. Among these, 54% had a deviation of the intergluteal fold and 35%, lumbosacral and/or perineal angioma. Perineal malformations were constant. Anal or rectal imperforation was present in 66% of the patients, recto-perineal, recto-vaginal or recto-urethral fistulas in 27.8%, and bladder extrophy in 6.2% (1 case).

Neurological deficits existed in 8 cases (50%). They were progressive in all 8, but obviously congenital in 4.

Four families and 56% of the children studied in our series presented with a family history of malformations in the lower lumbosacral region (Fig. 25). In 2 cases, the malformation was sacral agenesis. It seems therefore that an excess of anomalies exists in the parents of these patients compared with the estimated incidence of 22% for occult spina bifida in the general population [25]. Currarino syndrome is considered to have a dominant autosomal mode of inheritance with variable penetrance.

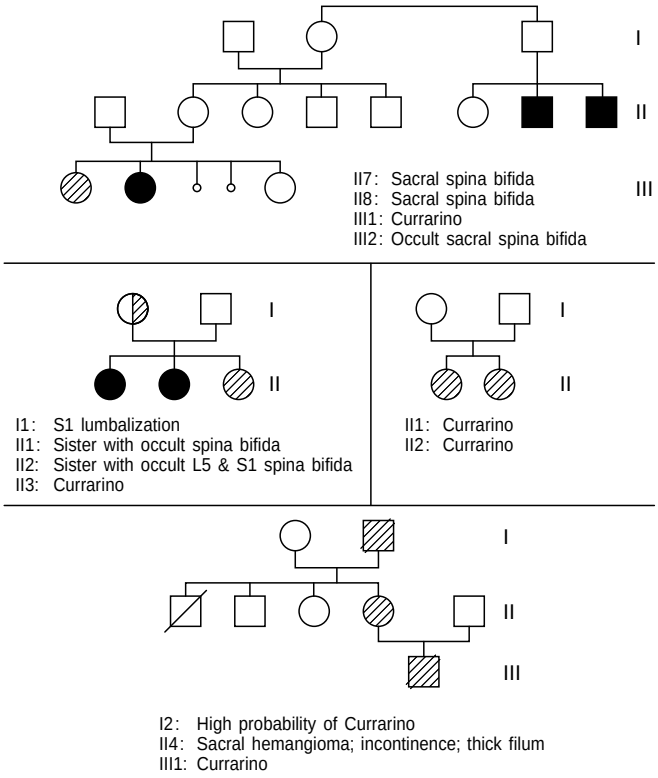


Fig. 25 Currarino syndrome. Family trees

Functional scores. To appreciate the functional and social repercussions of these malformations more easily and to allow rapid comparison between patients, we developed two functional scores, one for infants and one for older patients (Tables 15, 16). These scores take into account the major handicaps; motor, sensory, vesical and anal. The more disabling the handicap, the lower the score. These functional scores are simpler than those previously reported [6, 33] and differ widely from some others proposed in the neurosurgical literature [79, 138], which evaluate the neurological level of the deficits but not their functional repercussions. Normality was given a score of 5 for both motor and urinary functions, 4 for sensory function, and 3 for anal function.

Table 15 Personal NEM scores (children and adults)

	Motor	Sensation	Bladder	Anus
1	Wheelchair	Skin ulceration or amputation	Incontinence day and night	Incontinence
2	Major orthesis or two crutches	Pain	Nocturnal incontinence	Painful constipation
3	Minor or distal orthesis	Painless sensory deficit	Intermittent catheterization	Normal
4	Fatigue on walking	Normal	Dysuria or infections or stress incontinence	
5	Normal		Normal	

Table 16 Personal NEM scores (neonates and infants)

	Motor	Sensory	Bladder	Anus
1	Major deficit		Incontinence	Incontinence
2		Pain	Retention	
3	Club foot or atrophy or distal deficit			Normal
4		Normal	Dysuria or infections or stress incontinence	
5	Normal		Normal	

Handicaps caused by lipomas of the conus were more severe than those provoked by lipomas of the filum (Table 17).

Progressive neurological deficits. Progressive neurological deterioration has been well documented with lipomas of the filum as well as with lipomas of the conus. Many authors have emphasized that neurological deterioration can start at any age, including adulthood [60, 93, 108, 109, 119, 141] and old age [172]. Deterioration usually occurs slowly and insidiously [29, 32, 75, 79, 84, 93, 108, 141, 168], but rapid evolution and acute onset of symptoms have also been reported [53, 91, 108]. Among our patients, neurological deterioration was also insidious in the majority of cases, but was rapid in 20 patients (6%), including 3 infants, and acute in 8 (2.5%). Symptoms were genitosphincteral problems in 56% of cases, motor difficulties in 45%, pain in 14%, and bowel dysfunction in 12%.

Some authors have noted that aggravations can either follow sudden effort or situations leading to exaggerated bending of the back, or accompany pregnancy or the development of obesity [60, 75, 89, 108, 141, 168, 195]. The same findings were observed among our patients: 4 worsened following sudden and violent efforts, 1 after trauma (falling out of bed), 3 during development of obesity, 2 during pregnancy. These circumstances were constant in cases of sudden deterioration, and in half of the cases when rapidly evolving. Two youths thought their military service, and the physical exertion demanded of them at that time, had triggered their clinical deterioration.

The incidence of neurological deterioration in patients with lipomas is controversial. In the literature, the general impression is that deterioration is almost unavoidable with age: “uncertain number, perhaps the majority” [127]; “all will have neurological, urological, or orthopedic problems” [168]; “the risk of deterioration is high at all ages and increases with time” [47]; “a large number of children with an asymptomatic leptomyelo-lipoma will develop progressive neurological abnormalities” [32]; “we have no statistics available. In many studies, the risk appears to be at least 80%” [108]; “these patients deteriorate with increasing age” [79]; “the risk of deterioration is high at all ages and increases with age” [147].

In a previous publication that included data from the literature [142], we estimated the risk of deterioration at 56%. On reconsidering the problem, we noted that, in the present series, neurological worsening was clearly observed by the medical team in 35 patients (11.5% of the cases), but reported as clearly indisputable by 33 of the 161 symptomatic patients (20.4%).

Neurological imaging

Recent advances in prenatal ultrasonography and the onset of MR have greatly facilitated the diagnosis of lumbosacral lipomas. At present, both prenatal diagnosis of the malformation and postnatal correct anatomical analysis of it are possible.

Prenatal ultrasonography

The only two publications dealing with the prenatal diagnosis of lumbosacral lipomas are those of Aoki [8, 9], which concern 2 successive children in the same family. The prenatal diagnosis was made respectively at the 17th and the 26th week of gestation on the basis of the follow-

Table 17 Functional scores in patients with lipomas of the conus and with lipomas of the filum

Scores	Motor		Urinary		Sensory		Intestinal	
	Conus	Filum	Conus	Filum	Consus	Filum	Conus	Filum
5	68%	84%	59%	76%				
4	13%	11%	14%	18%	67%	86%		
3	13%	5%	4%	0%	20%	3%	78%	81%
2	2%	0%	4%	3%	11%	11%	11%	11%
1	4%	0%	19%	3%	2%	0%	11%	8%

ing association: lumbosacral spina bifida, anechogenic meningocele, overlying hyperechogenic skin and no hydrocephalus. Both the children were born with a lipomeningomyelocele.

In our series, 6 children born with a lipoma were diagnosed prenatally between the 17th and the 35th week of gestation as having lumbosacral dysraphism. In all of them, prenatal ultrasound revealed spina bifida associated with a subcutaneous hypoechogenic mass indicating a meningocele. In 1, the meningocele was separated into two by a longitudinal hyperechogenic septum. In all of them, hyperechogenic linear structures were visible within the meningoceles, suggesting the presence of either a low-lying spinal cord or roots (Fig. 26). None of these 6 fetuses had hydrocephalus or signs of Chiari malformation. The amniotic fluid was normal in all 6 cases.

In 4 fetuses, the meningocele was covered superficially by a thick hyperechogenic tissue. Because this resembled fat, a diagnosis of lipomyelomeningocele was made. In the 2 other fetuses the covering of the meningocele appeared to be normal skin, so that the prenatal diagnosis was one of meningocele. The absence of visible fat in these last 2 cases raises questions, since postnatally the thickness of the lipoma covering the meningocele was significant. This contradiction could be explained either by late prenatal growth of the lipoma or by the fact that lipomas could have different echographic behavior according to the amount of fibrotic tissue within the lipoma [135] or their water content level [21]. Postnatally, at surgery, the lesions were typical of a lipomeningomyelocele in 5 babies, and of a lipomyelocystocele in 1. The prenatal ultrasonography of this last patient differed from the others in that the meningocele was apparently double (Fig. 27). In no case did prenatal MRI provide more information than the echographies.

Postnatal imaging

Plain X-rays

Plain X-rays were routine in our series. However, lack of ossification in neonates and infants made their interpretation difficult, especially concerning the sacrum, and this raises the question of their usefulness in this age range. Even if they do not demonstrate the existence of a spina bifida very well, in our opinion they must remain part of the routine check-up in searching for other possible spinal abnormalities, such as diastematomyelia, abnormal vertebral segmentation, abnormal spinal curves, and sacral agenesis.

Spinal echography

Ultrasonography is the screening examination of choice up to the age of 6 months [167] because it is cheap and noninvasive [37, 104, 136, 151, 167, 199]. Our attitude would be to recommend ultrasonography as a first investigation in asymptomatic infants with lumbosacral cutaneous stigmata. In the presence of deficits, the usefulness of echography is, however, less than in their absence, since MRI will be necessary in all cases.

Ten children in our series were primarily investigated with echography. In 2 cases, the lipoma involved the filum, and in 8 the conus. In all but 1, the cord termination was abnormally low (Fig. 28). In all patients, the lipoma was visible as an intraspinal, hyperechogenic, homogeneous and well-demarcated mass. In our cases, as in those of Naidich et al. [136], lipomas were more echogenic than was epidural fat. In the literature, there is only 1 case of a false-negative echogram. As in the prenatal period, the question is why lipomas may have different echogenicity [21, 135].

Magnetic resonance imaging

MRI is considered by all as the necessary investigation prior to surgery [5, 15, 34, 70, 103, 105, 134, 140, 159, 179, 186, 194]. Since 1984, all our patients have had preoperative MRI including sagittal and axial views in T1- and T2-weighted images to ensure correct analysis of the relative positions of lipoma and spinal cord, to foresee, if possible, the type of insertion of the lipoma on the cord and hence the situation of the roots with regard to the lipoma, and to look for the possible coexistence of lipomyelocele, meningocele, syringomyelia or other malformations. In addition, for the past 2 years a routine dynamic MRI has been performed to evaluate the mobility of the

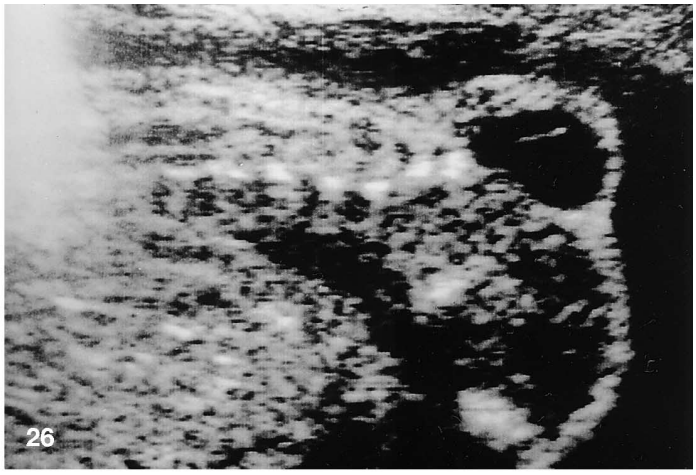
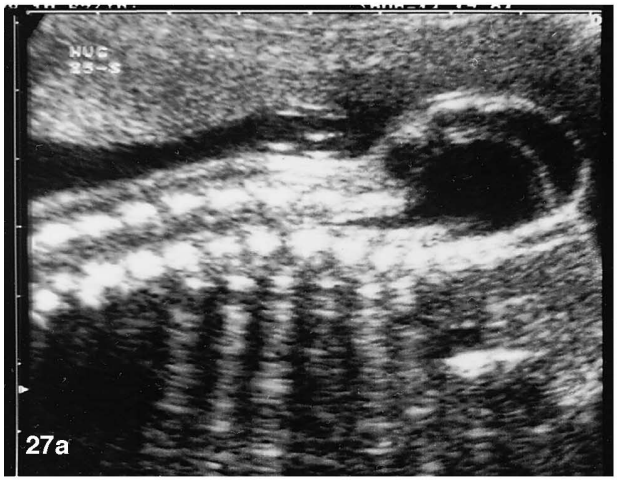


Fig. 26 Prenatal ultrasonography, sagittal view, showing a lumbosacral meningocele separated into two by a longitudinal hyperechogenic septum. At operation, the malformation was diagnosed as a lipomyelocystocele and the longitudinal septum, as the structure separating the syrinx from the subarachnoid spaces



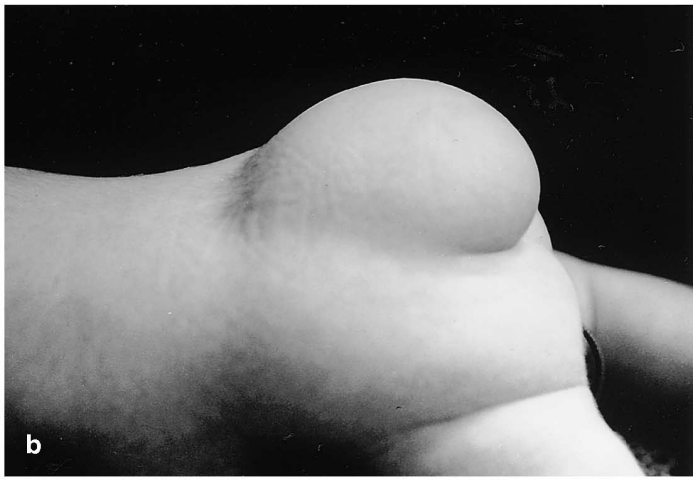
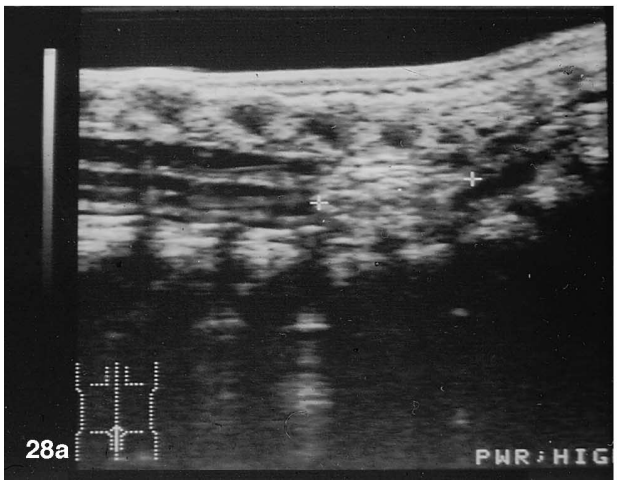


Fig. 27 **a** Prenatal ultrasonography, sagittal view, showing a lumbosacral meningocele superficially limited by an abnormally thick and hyperechogenic covering. A linear hyperechogenic structure within the meningocele indicates the presence of either a low-lying cord or roots. The prenatal diagnosis was that of a lipomyelomeningocele. **b** Same patient a few days after delivery. The lipoma, much larger than prenatally, actually filled the whole of the prenatal meningocele



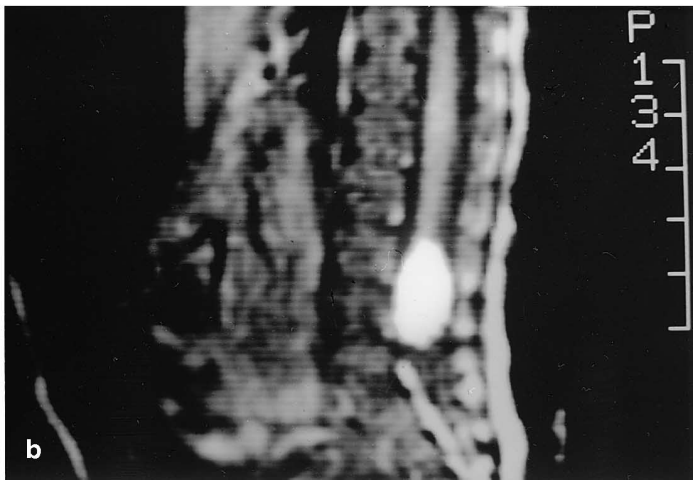


Fig. 28 **a** Postnatal echography, sagittal view. Presence of a hyperechogenic mass occupying the lumbosacral canal and of a low-lying spinal cord ending at the upper pole of the mass. **b** Postnatal MRI, same patient. Tethered and low-lying spinal cord anchored to a lumbosacral lipoma

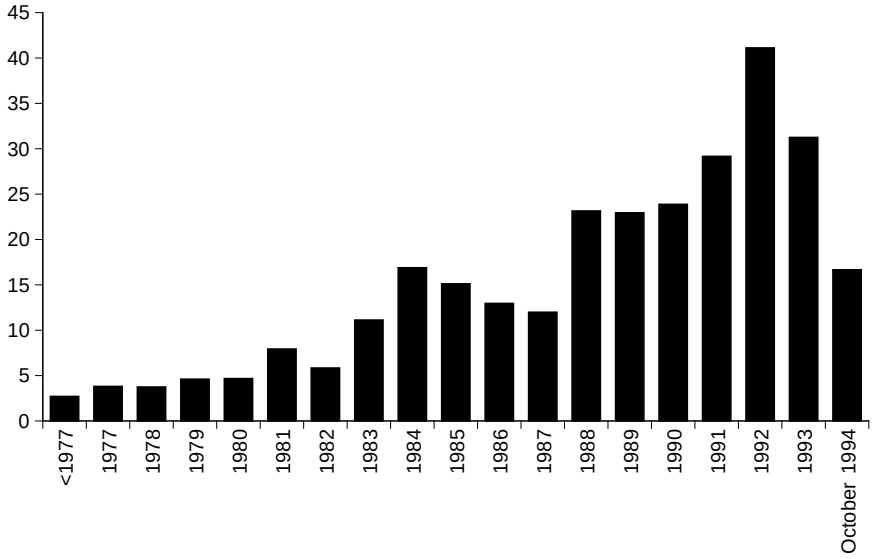


Fig. 29 Number of surgical interventions per year

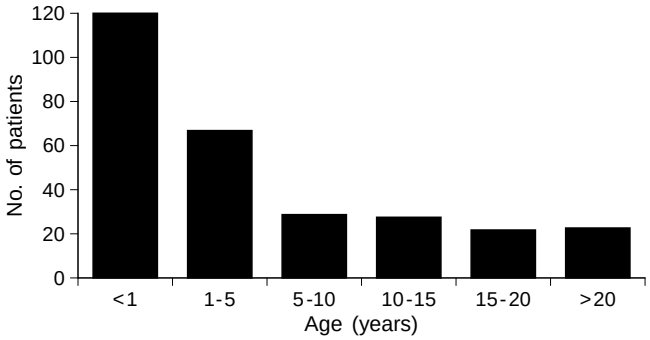


Fig. 30 Age at the time of surgery

lumbar cord [35]. Recently, Johnson and Levy emphasized the usefulness of dynamic MRI by studying the mobility of the cord at its cervical level in the tethered cord syndrome [90].

Surgical treatment

The proportion of operated patients has regularly increased over the years (Fig. 29). The majority of our patients were under the age of 1 at time of surgery (Fig. 30).

Principles

As for the vast majority of neurosurgeons, our objectives have been to free and decompress the cord when necessary, while sparing functional nervous tissue and preventing retethering. To achieve these aims, our surgical goals have been to remove as much lipoma as possible, to divide adhesions, and enlarge the dural sac before closing.

Operations were conducted under optic magnification, using the ultrasonic aspirator (CUSA) to remove lipomas, as advocated by many [43, 47, 71, 79, 88, 126, 175], and intraoperative monitoring to recognize the functional nerves when necessary [148]. Responses to stimulation of unidentified structures were sought at three levels: lower limbs by mere visual inspection, bladder by intravesical pressure recordings, posterior funiculi of the cord, a few centimeters above the upper limit of lipomas, by somatosensory evoked potential recordings (SSEP). Nonfunctional nerves, or non-neural structures mimicking normal nerves were sacrificed when necessary. Many publications have also insisted upon the usefulness of intraoperative detection of functional nervous structures by using SSEP recordings [75, 146, 170] or urodynamic studies [170], as we did. Some others proposed more sophisticated techniques of recordings using electromyography of the lower limbs [170], electromyography of the bladder [170], or of the anal [86–88], or intrarectal pressure [76, 81, 141, 163]. All these techniques have, however, the common disadvantage of not allowing permanent monitoring of the surgical procedure. To obtain such control, we recorded SSEP in response to continuous stimulation of the pudendal nerves. Unfortunately, this procedure did not give us reliable information, probably because of the immaturity of the nervous system in young children.

Technique

The technique used depended upon the type of lipoma.

Lipomas of the filum

Lipomatous filums were cut or removed, depending upon the site and extension of the fatty infiltration. The surgical

approach to the filum was easy in all cases. Subcutaneous lipoma was usually absent, and the lumbosacral-sacral aponeurosis intact. In 70% of cases the dural sac was normal. In the other 30%, it was moderately agenetic. Once the dura was open, care was taken not to injure the sacral roots, which were often attached by pronounced adhesions to the pia mater covering the lipomatous filum. In 1 case only, sacral roots were subpial and intralipomatous, making removal of the lipomatous filum impossible. A single similar case was reported in the literature [30]. Another rare difficulty consisted in the extreme shortness of the filum, and consequently in recognizing the filum from the conus. As emphasized by Hendrick et al. [76], the cord was actually always accompanied by a large satellite vein that indicated where cutting was possible or not possible. The dural sac, following resection or cutting of the lipoma, was not enlarged in any case reported.

Lipomas of the conus

In the case of lipomas of the conus, surgery was much more difficult. The skin incision, always vertical and median, was curved horizontally in its lower part to remain as far as possible from the anal region when lipomas were situated low down. In similar cases, some authors have proposed horizontal [18] or W-shaped [75] incisions. When the subcutaneous lipoma was voluminous, the skin incision was split in two at the top of the cutaneous lump to allow resection of both the excess skin and the lipoma. The subcutaneous lipoma was totally removed, except in the last 12 cases of the series, where a major part was left in situ to avoid the creation of a large subcutaneous dead space in which the CSF could collect postoperatively.

Laminectomy of the lowest normal laminae was often necessary, either to expose the upper limit of the intradural lipoma correctly or to diminish the lumbar curve which, in some cases, was partly responsible for tethering of the cord.

The dural incision was made rostro-caudally and circumferentially at the point where the lipoma penetrated the dural sheet. At this level, adhesions between lipoma, dura and spinal cord were frequently severe and had to be carefully divided to expose the underlying roots. Removal of the intradural lipoma was easy when its cord insertion was caudal or dorsal, and posterior roots were extra-lipomatous and easily recognizable. In contrast, it was difficult when its insertion was dorso-lateral or lateral, roots were malformed, either short, aberrant or intralipomatous, and dura largely missing, replaced by fibro-muscular bundles. Severe adhesions also often complicated surgery badly by hiding nerve roots from the view of the surgeon (Fig. 31) or by mimicking nerve roots. In such cases, dissection was often so difficult that, despite the help of intraoperative monitoring, surgery was stopped before correct freeing of the cord was achieved (Fig. 32).

Closure with enlargement of the dural sac, as recommended by many [1, 26, 47, 79, 82, 185] to limit risks of postoperative adhesions, and hence of retethering, was performed routinely in the last 90 patients of the series (33%). This procedure was difficult when the dura was largely missing laterally. In such cases, the repair had to be fixed to the paraspinal muscles or aponeurosis making tight sutures impossible. The literature reports that many types of materials have been used for dural repair, such as fascia [71, 79], lyophilized dura [1, 79], amniotic membrane, plain Vicryl, or Vicryl reinforced with Dacron, Dacron, Silastic, Medpor [197] or Gore-Tex [82, 98]. In our series, the material used were lyophilized dura ($n = 37$), amniotic membrane ($n = 6$), autologous plasty ($n = 36$), and synthetic plasty ($n = 21$). In the vast majority of our cases autologous material led to severe postoperative adhesions, which were visible not only on postoperative MRI (Fig. 33), but also intraoperatively at reoperation. Lyophilized dura had the same drawback, and involved the further major disadvantage of possibly inoculating Kreutzfeld-Jakob disease. Dacron gave us apparently satisfactory results, but has been reported as causing postoperative hemorrhages and severe adhesions [26, 98]. Gore-Tex and Silastic were satisfactory in all cases (Fig. 34), both in our experience and in that of Boop and Chadduck [26].

The possibility of correct musculo-aponeurotic closure depended upon the site of the spina bifida. When lumbar, closure has always been tight, sometimes attained by releasing and mobilizing the lumbodorsal fascia towards the midline, the technical choice depending upon the width of the spina bifida. When sacral, contrary to Zide's statements [197], closure has always been incomplete and not tight in our hands, especially in the case of sacral agenesis. At that level, muscles and aponeurosis lie too far away from the midline to be correctly mobilized towards the midline. For that reason, we felt that surgery should avoid creating a subcutaneous dead space by leaving the major part of the subcutaneous lipoma. This was done in our last 12 consecutive patients and was successful.

Three questions still remain unanswered:

1. How much lipoma should be removed? Opinions vary from minimal [164] to partial [47, 79, 126] or to complete [175] removal. Our policy was to do as near-complete a removal as possible. This could be achieved in 167 of the 252 lipomas of the conus (66%). Of the remaining 85 cases (34%), removal was partial in 81 and minimal in 4. There is no doubt that "total" removal in an attempt to reach the fibrotic interface between lipoma and cord carries the intrinsic risk of injuring the posterior columns. This happened in 3 of our patients, in 2 as the result of first surgery, and in 1 after reoperation. These 3 patients complained postoperatively of insensitive bladder, at least during the first few postoperative weeks. For that reason, we no longer advocate as complete a removal as we did in the past, especially as we found no correlation between postoperative results and the extent of removal.

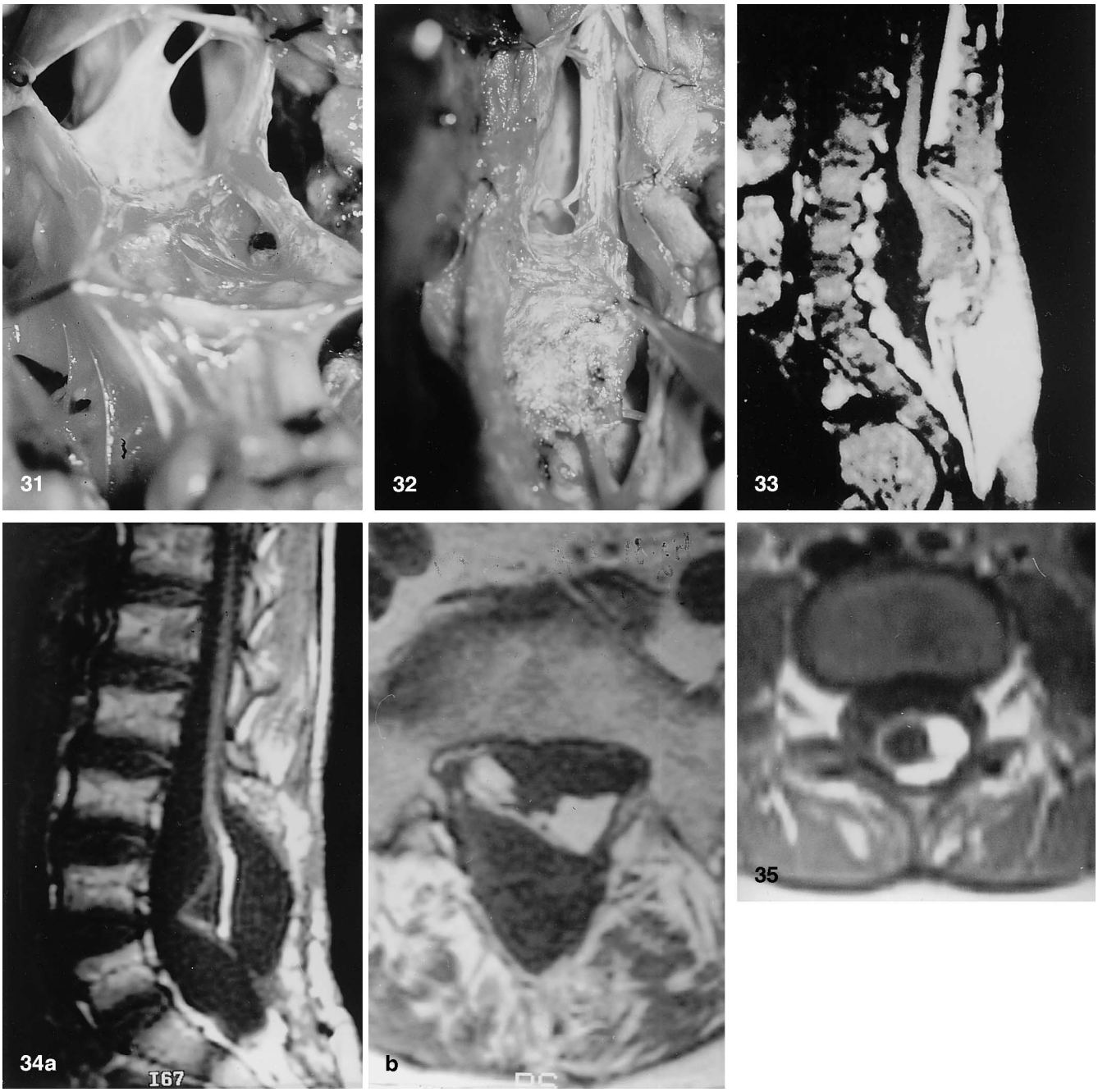


Fig. 31 Operative view. The dura is open and the lipoma still unremoved. Note the severity of the adhesions between the lipoma, the dura and the spinal cord, not only at the level of the medullary insertion of the lipoma but also above it

Fig. 32 Operative view after the resection of a lipoma. The cord remains attached to left aspect of the dura owing to severe adhesions that hide the roots on the same side

Fig. 33 Postoperative MRI, T1 relaxation time, sagittal view, showing severe adhesions between the posterior aspect of the cord and the lyophilized dura used to enlarge the dural sac

Fig. 34 Postoperative MRI, T1 relaxation time, sagittal median (**a**) and axial (**b**) views. The spinal cord is free within the mega-cul de sac. The dural sac has been enlarged by means of a Gore-Tex duroplasty

Fig. 35 MRI, T1 relaxation time, axial view: syringomyelia partly confined by a lipoma. Resection of the lipoma will lead to opening of the syrinx

2. Should the edges of the placode be closed? This was advocated by McLone et al. [127] to limit the risk of postoperative reattachment. Closure of the placode was performed in 17% of our cases. Limited postoperative adhesions were observed in the 3 patients who underwent reoperation. Adhesions were found only on the midline at the level of the stitches, making reoperations much easier and safer than in absence of closure of the placode. This technique however, did not seem to protect patients from late postoperative deterioration.

3. Should syringomyelic cavities be drained? Choux et al. [47], Iskandar et al. [83] and Hoffman et al. [79] advocated myelotomy and/or drainage in the case of severe syringomyelias. In our series, syringomyelic cavities were not treated. Cavities were only opened when lipomas, as in Fig. 35, were in direct contact with them. Contrary to Iskandar's findings [83], there was no correlation in our study between patients' pre- or postoperative clinical status and the presence or size of a syringomyelic cavity. Furthermore, many syringomyelias shrank postoperatively without being drained. These data made us conclude that treatment of syringomyelias was not necessary in the great majority of cases.

Surgical complications

The literature shows that it has been the practice to study lipomas of the filum along with those of the conus and even, in some series, with other types of dysraphism, as if these malformations were comparable. In this study, we analyzed lipomas of the filum and of the conus separately because, as emphasized above, they are entirely different, not only clinically and anatomically, but also from a surgical point of view. Therefore, it is difficult to compare our therapeutic results with those in the literature.

Lipomas of the filum

Of 39 lipomas of the filum in our series, 38 were operated on. In 37 cases, the lipomatous filum was cut or removed as expected. In 1 case (2.6%) this could not be achieved owing to the presence of lipomatous roots.

There was no case of operative mortality, and no local, general or neurological complications. These results seem to correspond with those in the literature, most reported complications apparently being secondary to surgery on lipomas of the conus.

Lipomas of the conus

Of 264 lipomas of the conus in the series, 253 were operated on. Decompression and/or untethering of the nervous system were correctly achieved in 202 of these 253 cases (80%). In the remaining 51 patients (20%), surgery was stopped owing to the complexity of the malformations.

Operative mortality

There was no operative mortality in this series. This is in accordance with the literature, where it averages 0.2%.

General complications

Not mentioned in the literature, general complications were rare in our series (0.7%): 1 patient developed septicemia and 1, pulmonary embolism.

Local complications

Local complications affected almost 20% of our patients (Table 18).

In the literature, they ranged from 0% [47, 175] to 56% [198], and they were estimated by McLone [125] at an average of 25%. In all series, infections were mainly local due to the proximity of the wound to the anal region [32, 85, 93, 137].

Subcutaneous pseudo-meningoceles were frequent, especially in the case of sacral lipomas [197], and were responsible for wound breakdown, CSF leak, and infection [126, 197]. In our patients, treatment has usually been long and tedious, necessitating repeated lumbar drainage, compressive dressings and, in more than 8% of cases, additional surgery (Table 19). Various procedures have been proposed to minimize their incidence, all aiming to close

Table 18 Postoperative local complications in 58 patients: NEM series ($n = 291$)

Complication	*n*	%
Meningocele	43	47.8
Subcutaneous infection	17	18.9
CSF leak	13	14.4
Wound non-union	13	14.4
Meningitis	2	2.2
Skin necrosis	2	2.2
Total	90	100.0

Table 19 Reoperations for local complications

NEM series ($n = 291$)	*n*	%
Wound repair	19	6.5
External lumbar drainage	1	1.3
Lumbo-peritoneal shunt	4	0.3
Total	24	8.2

the dura and the musculo-aponeurotic layers better. As already stated, these techniques were ineffective in our hands. The only technique that prevented postoperative meningoceles was leaving the major part of the subcutaneous lipoma in situ.

Neurological complications

All our patients had postoperative EMG and urodynamic studies. We considered as aggravated by surgery (1) patients who postoperatively presented with a new symptom or a symptom that was worse than preoperatively, even when other preexisting deficits improved or disappeared following surgery, and (2) babies in whom postoperative EMGs or urodynamic studies were worse than preoperatively, even in the absence of clinically detectable changes. Thirty-two patients were postoperatively aggravated (10.9%), 22 transiently (7.5%), and 10 permanently (3.4%).

Transient deterioration. Table 20 indicates the types and incidences of transient aggravations. All completely regressed in a maximum of 6 weeks, while EMGs and results of urodynamic studies always returned to their preoperative status. The incidence of such transient deteriorations has never been mentioned in the literature, but their possibility was reported by Dubowitz et al. [53], James et al. [85], Linder et al. [117], and Swanson and Barnett [178].

Permanent deterioration. Permanent deterioration occurred in 10 of the 253 patients (3.9%) operated on for lipoma of the conus. Their type and incidence are shown in Table 20. Sphincteral disturbances were more frequent than others, and were explained by bladder hyposensitivity rather than by bladder paralysis. Five patients had only postoperative urinary complications, 3 had motor and/or sensory deficits in a lower limb, and 2 had both types of disorders. The postoperative worsening was severe in 3 patients, but mild and only slightly handicapping in 7. As an example, in these 3 cases, preoperative frequency of micturition with urinary leaks transformed into urinary retention but continence was maintained with self-catheterization.

In the literature, it is difficult to assess the postoperative neurological risk correctly. More than half of the articles do not mention postoperative complications, and many others either group all types of lipomas, and even sometimes all types of spinal dysraphisms, in the same analysis or present the total number of their immediate and delayed postoperative aggravations. Reported rates of postoperative deteriorations range from 0% [7, 32, 44, 53, 75, 79, 101, 175] to 4% [47, 93, 126, 136] of postoperative aggravations.

Risk factors. In our study, 15 parameters were entered into a statistical study to search for risk factors: date of surgery, patients' age and sex, clinical presentation, MRI aspects of the cord and lipoma, associated malformations. Only 3 parameters were statistically correlated with the onset of postoperative complications: date of surgery, anatomical type of lipomas (filum vs conus), and associated adhesions and/or radicular malformations. There were 10 cases of postoperative complications, 7 following surgery performed before 1988, 2 following surgery done between 1988 and 1991, and 1 following surgery done after 1991. The 10 aggravations concerned lipomas of the conus, and, of these 10, severe adhesions were described in all and severe root abnormalities in 6. We can therefore conclude from this study that the surgeon's level of experience and the anatomical complexity were the only prognostic factors. These conclusions are not surprising. They were partly confirmed by Choux [47], who reported that 3 of the 4 cases of postoperative aggravation in his series of 96 patients occurred in patients with severe root abnormalities [149].

Mechanisms of deterioration. Most deteriorations in our series probably resulted from lesions of undetected functional roots or of the posterior columns of the cord. Some, however, were possibly the result of vascular complications. This is illustrated by the following observation: a 49-year-old man was operated on in 1991 for severely progressive motor and sphincter deficits involving L4 to S4. The lipoma was compressive, filling the entire dural sac up to L4. Surgery was accompanied by hemorrhage. After a 7-h dissection, only half the lipoma was removed when surgery was stopped. The patient woke up with flaccid paraplegia and soon after developed encephalopathy, renal failure and pancreatitis associated with severe myoglobulinemia. These latter complications progressively cleared up, but the paraplegia remained unchanged. On examination, the upper sensory level was found to be T8. The discrepancy between this sensitive level and the lumbosacral localization of the lipoma strongly suggests a vascular complication attributable to abnormal spinal cord vascularization.

Table 20 Neurological complications seen immediately after surgery

	Transient		Permanent	
	n	%	*n*	%
Urinary retention	16	5.5	4	1.4
Incontinence	7	2	6	2.1
Anal paralysis	1	0.3	0	0
Motor deficit	14	4.8	6	2.1
Superficial sensory deficit	5	1.7	3	1
Deep sensory deficit	4	1.4	2	0.7
Total[a] (patients)	22	7.6	10	3.4

[a] Some patients had multiple complications

Surgical results

To date, the results of lipoma surgery remain controversial and difficult to analyze. In the literature, follow-up was frequently short, at less than 2 years [59, 75, 111, 126, 127], or not mentioned [32, 47, 108, 109], and the pathology reported covered all causes of tethered cord syndrome [7, 20, 36, 58, 69, 95, 97, 101, 153]. In addition, many analyses did not take into account the patients' preoperative clinical status [79] or the length of follow-up [7].

To clarify the subject, we present our results according to (1) anatomical type of lipomas (conus or filum), (2) preoperative status, (3) degree of lipoma removal and freeing of the cord, (4) age at surgery, and finally, (5) length of follow-up. The follow-up results have been studied at 1 year postoperatively and at the maximum follow-up for each patient. The average maximum follow-up in this series was 6.3 years. Furthermore, a subgroup of 93 patients followed up for more than 5 years has been individualized to give better idea of the long-term results (range from 5 to 23 years; average 8.7 years; median 8 years). Comparison with the literature was again difficult. Only eight series provided follow-up for longer than 4 years [38, 79, 93, 94, 111, 116, 185, 196]. In our study, patients were classed as being impoved or cured when their preoperative deficits improved or disappeared, provided however that no other deficit appeared or worsened with time, in which case they were classed as having deteriorated.

After 1 year

Lipomas of the filum

As indicated in Table 21, no patient deteriorated with time, and 9 of the 17 (53%) who were preoperatively symptomatic were cured or improved. All types of deficits regressed. Of the others, 2 patients stopped deteriorating and 8 remained stable.

In summary, surgical results were good in all cases. In around 65% of the patients, surgery resulted in clinical improvement.

Lipomas of the conus

As indicated by Table 21, 20 of the 253 operated patients deteriorated (7.9%): 7 of the 109 without preoperative symptoms, and 13 of the 144 with preoperative deficits. Of these 20 deteriorations, 10 were secondary to surgery and affected patients who were preoperatively asymptomatic in 2 cases and symptomatic in 8. A total of 103 patients (71.5%) were either cured ($n = 26$) or improved ($n = 72$), or stopped deteriorating ($n = 31$), regardless of the type of their deficits.

In summary, 6.4% of the asymptomatic patients and 9% of those with preoperative deficits worsened. Of the remaining patients, 72% of the symptomatic patients improved or stopped worsening and 19.1% stabilized. As for lipomas of the filum, all types of preoperative troubles regressed postoperatively.

At maximum follow-up

Lipomas of the filum

Of 21 preoperatively asymptomatic patients, 20 (95.2%) remained asymptomatic (Table 22). The 1 patient who developed symptoms showed only moderate deterioration, complaining of a slightly disabling dysuria related to a hyperactive bladder.

Table 21 Clinical status 1 year after surgery

Preoperative status / Postoperative status ($n = 291$)	Normal ($n = 130$)				Abnormal ($n = 161$)			
	Filum		Conus		Filum		Conus	
	$n = 21$	%	$n = 109$	%	$n = 17$	%	$n = 144$	%
Normal	21	100	102	93.6	7	41.2	26	18.1
Improved					2[a]	11.8	46[a]	31.9
Motor					2	5.9	22	15.3
Sphincter					2	5.9	33	22.9
Both					2	11.8	8	5.6
Stabilized					8 (2)[b]	47	59 (31)[b]	41
Worse	0	0	7[a]	6.4	0	0	13[a]	9
Motor			5	4.6			8	5.6
Sphincter			4	3.7			11	7.6
Both			2	1.8			6	4.2

[a] Number of patients; [b] Patients with deficits evolving preoperatively

Table 22 Clinical status at maximal follow-up after surgery

Preoperative status / Postoperative status ($n = 291$)	Normal ($n = 130$)				Abnormal ($n = 161$)			
	Filum		Conus		Filum		Conus	
	$n = 21$	%	$n = 109$	%	$n = 17$	%	$n = 144$	%
Normal	20	95.2	83	76.1	6	35.3	26	18.1
Improved					2[a]	11.8	46[a]	31.9
Motor					1	5.9	21	14.6
Sphincter					2	11.8	25	17.4
Both					1	5.9	0	0
Stabilized					9 (2)[b]	52.9	43	29.9
Worse	1[a]	4.8	26[a]	23.9	0	0	29[a]	20.1
Motor	0	0	12	11			17	11.8
Sphincter	1	4.8	18	16.5			16	11.1
Both	0	0	4	3.7			4	2.8

[a] Number of patients; [b] Patients with deficits evolving preoperatively

In symptomatic patients, surgery was beneficial in at least 10 (58.8%) of the 17 whose deficits either regressed or stopped evolving. The 9 others remained unchanged. In summary, surgical results at maximum follow-up remained as good as at 1 year.

Lipomas of the conus

With lipoma of the conus, 26 of the 109 preoperatively asymptomatic patients had deficits (23.9%), and 83 were still symptom-free (76.1%). Since, the 1-year postoperative follow-up, 16 patients (17.5%) had worsened (Table 22).

Among the group with preoperative deficits, 72 were still cured or improved (50%), 43 were unchanged (29.9%), and 29 were worse (20.1%). Compared with the results at 1 year, 16 additional patients had developed new deficits or their previous deficits had worsened (11.1%).

In summary, at maximum follow-up, results had slightly deteriorated with time in all groups of patients. However, 50% of them were still greatly improved by surgery.

At more than 5 years of follow-up

The 93 patients followed up for longer than 5 years have all been reviewed in the last 2 years preceding this study. Ten had a lipoma of the filum and 83 a lipoma of the conus.

Lipomas of the filum

As indicated in Table 23, results were unchanged. Among the 7 patients with no preoperative deficits, 6 were still symptom free (85.7%) and 1 was slightly worse than before surgery. This patient is the 1 already mentioned at 5 years postoperatively. All 3 patients who were symptomatic at time of surgery were cured when last reviewed.

In summary, the good immediate postoperative results were maintained with time, whether patients were preoperatively symptomatic or not.

Lipomas of the conus

Of the 32 patients with no preoperative deficits, 17 were still symptom free (53.1%), but 15 had deficits (46.9%). Thus, of these patients, who were symptom free before surgery and operated on prophylactically, 93.6% were still neurologically normal 1 year after surgery, but only 53.1% at an average of 8.7 years after operation (Table 23).

Of the 51 patients with preoperative deficits, 31 were still improved (60.7%): 7 were cured, 20 improved, and 4 had apparently stabilized (Table 23). The long-term outcome of preoperatively symptomatic patients depended upon the immediately postoperative functional results. The better these results, the better the long-term neurological status. Most of those who were clearly improved 1 year after surgery remained improved. Conversely, most of those who deteriorated with time either were not improved by surgery or only stabilized (Fig. 36).

In summary, very long-term surgical results were much less favorable for lipomas of the conus than for those of the filum. Overall, in both groups of lesions, all but 1 of the long-term deteriorations occurred in patients with lipomas of the conus. More than 60% of the patients with such lipomas benefited from surgery, while around 30% of those with preoperative deficits deteriorated with time, as did 47% of those operated on in the absence of neurological deficit.

Table 23 Clinical status of patients with more than 5 years of follow-up

Preoperative status / Postoperative status ($n=93$)	Normal ($n=39$)				Abnormal ($n=54$)			
	Filum		Conus		Filum		Conus	
	$n=7$	%	$n=32$	%	$n=3$	%	$n=51$	%
Normal	6	85.7	17	53.1	3	100	7	13.7
Improved					0	0	20[a]	39.2
Motor							15	29.4
Sphincter							12	23.5
Both							7	13.7
Stabilized					0	0	9 (4)[b]	17.6
Worse	1[a]	14.3	15[a]	46.9	0	0	15[a]	29.4
Motor	0	0	6	18.8			7	13.7
Sphincter	1	14.3	14	43.8			10	19.6
Both	0	0	3	9.4			2	3.9

[a] Number of patients; [b] Patients with deficits evolving preoperatively

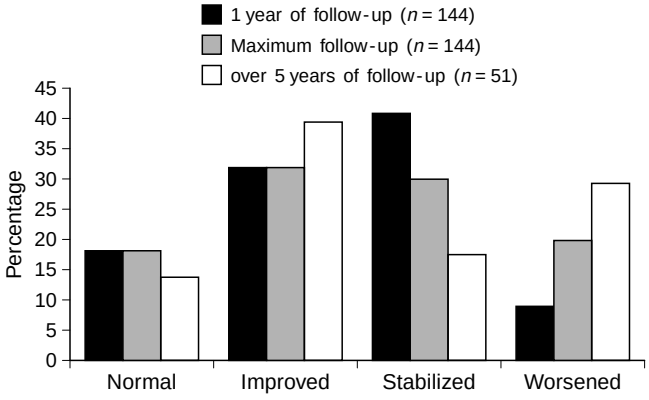


Fig. 36 Postoperative clinical outcome in patients with symptomatic lipomas of the conus

The functional loss in these patients was severe, an average of 3.8 points in our functional score. The sphincter function was the most affected (1.8 points), followed by the sensory function (0.9 points), the motor function (0.6 points), and the bowel function (0.4 points).

Prognostic factors

A search was undertaken for prognostic factors. Five parameters have been studied: anatomical type of lipomas (filum or conus), quality of surgery, age at surgery, and malformation complex.

Anatomical type

There was a highly significant correlation between the anatomical type of lipoma (filum/conus) and the long-term outcome. When preoperatively symptomatic and asymptomatic patients were grouped together, rates of deterioration at 5 years or more following surgery were 10% for lipomas of the filum and 36.1% for lipomas of the conus.

Quality of surgery

In the group of preoperatively asymptomatic patients, the extent of spinal cord decompression and untethering was statistically significantly correlated only with long-term prognosis ($P=0.0001$) (Table 24). This was not the case in patients who were symptomatic at the time of surgery.

Age at surgery

Contrary to what has been reported in the literature [11, 59, 97, 116], there was no difference in the long-term outcome between patients operated on before and after 1 year of age. Confirming some previous observations [14, 53, 111, 114], adults in our series improved as well as children.

Malformation complex

Sacral agenesis and perineal or visceral malformations were not correlated with prognosis. In summary, at present, the long-term prognosis for lipomas of the conus cannot be foreseen preoperatively.

Postoperative evolution of the functional status

In our experience, apart from scoliosis or club foot, all other types of neurological deficits were improved by surgery.

Table 24 Clinical status at maximum follow-up related to quality of surgery (lipomas of the conus)

Preoperative status / Postoperative status ($n=233$)[a]	Normal ($n=93$)				Abnormal ($n=140$)			
	Satisfactory		Unsatisfactory		Satisfactory		Unsatisfactory	
	$n=65$	%	$n=28$	%	$n=89$	%	$n=51$	%
Normal	54	83.1	14	50	12	13.5	7	13.7
Improved					43[b]	48.3	23[b]	45.1
Motor					21	23.6	10	19.6
Sphincter					22	24.7	13	25.5
Both					0	0	0	0
Stabilized					31	34.8	15	29.4
Worse	11[b]	16.9	14[b]	50	15[b]	16.9	13[b]	25.5
Motor	3	4.6	5	17.9	5	5.6	8	15.7
Sphincter	8	12.3	12	42.9	10	11.2	6	11.8
Both	0	0	3	10.7	0	0	1	2

[a] Number of patients; [b] For 20 patients no information was available

Pain, as in most other publications [12, 91, 117, 137], proved to be the most easily and rapidly curable symptom. Response of sphincter disturbances to surgery was obvious in many cases. However, for some others, the influence of surgery was more difficult to assess because after surgery the patients were managed medically in urological and physiotherapy units.

Resistance of spinal and foot deformities to surgery could be easily explained. Scoliosis, in the vast majority of our cases, was the consequence of either vertebral malformations or asymmetrical shortening of the limbs, and not directly related to neuronal dysfunction. This seems to be in accordance with the literature, which includes only one publication reporting on postoperative regression or stabilization of scoliosis [93]. With respect to club foot, there is a natural tendency for it to become constantly worse during growth owing to the pre-existing muscular imbalance at the level of the sole [84, 175] and to stabilize later once growth has finished. This is also a consistent finding, since James et al. [84] presented the only pediatric observation of a foot malformation that regressed postoperatively. However, if neurosurgery has almost no direct effect on the club foot, freeing and decompression of the cord must be performed before the orthopedic correction of foot malformations.

Comparison with the literature

The general opinion is summarized best by Anderson [7], who wrote in 1968: “most authors have concluded that operation controls the progression of symptoms; improvement is described in at least half of the patients and surgical morbidity is low.” In addition, as shown in Tables 25–27, which summarize data from the literature, the other four major results are that: (1) lipomas of the filum had a better prognosis than lipomas of the conus [91, 117], (2) all types of deficit regressed or stabilized postoperatively, the most resistant, however, being the sphincter deficits [93, 164, 196], (3) surgery on asymptomatic patients prevented them from further deterioration [110, 111, 116], and (4) satisfactory results immediately after operation are maintained over time [111, 116, 196]. These conclusions do not completely agree with ours.

Similarities

We agree with most of what has been written on symptomatic and asymptomatic lipomas of the filum, and on symptomatic lipomas of the conus.

For lipomas of the filum, our results were excellent and were maintained over time. For symptomatic lipomas of the conus, surgery was indisputably effective. Although long-term results were much better in patients who improved immediately than in those who were only stabilized, at least 50% of the patients benefited from surgery. All types of neurological deficits improved, pain more than other symptoms [114], whatever the patient’s age at referral [7, 126].

Differences

Differences, between the literature and our series concentrate on the long-term prognosis of asymptomatic lipomas of the conus. It is generally accepted, mainly since the publication of Bruce and Schut [32], that surgery gives the best guarantee that asymptomatic children will remain so. These conclusions are in striking contrast with ours: almost 50% of asymptomatic patients in our series developed deficits in the long term. This divergence could be

Table 25 Postoperative results from the literature

Reference	Year	Anatomical type		Follow-up	Asymptomatic		Symptomatic			Not indicated		
		Filum	Conus		Stabilized	Worse	Improved	Stabilized	Worse	Improved	Stabilized	Worse
[111]	1967	6	20	1–6 years	100%		52.9%	41.1%	6%			
[157]	1971						46.6%	46.6%	6.8%			
[185]	1973			11 years			5.1%	76.9%		4.5%	79.5%	16%
[155]	1974						71%	21%	8%			
[7]	1975	18	21							0%	100%	0%
[189]	1976									50%	50%	0%
[32]	1979									35%	59.5%	5.5%
[145]	1979									71%	15%	14%
[85]	1994									17%	73%	13%
[44]	1995									0%	100%	0%
[127]	1990-1									0%	76%	14%
[85]	1985									9.5%	87%	2.5%
[109]	1988	20	38		95%		73.9%					
[116]	1987			54 months	80%	20%	28%	32%	20%			
[175]	1988									27%	73%	0%
[119]	1990	4		22 months						3 cases		1 case
[193]	1993		19	2–4 years						15.7%	84.3%	0%
[38]	1995	10	90	5 years	93%	7%			27%			

Table 26 Postoperative results from the literature: filum vs conus

Reference	Year	Anatomical type		Follow-up	Asymptomatic		Symptomatic			Not indicated		
		Filum	Conus		Stabilized	Worse	Improved	Stabilized	Worse	Improved	Stabilized	Worse
[91]	1956	6					100%	0%	0%			
[117]	1982		21	19 months						24%	71%	5%
[193]	1993	15		19 months						47%	53%	0%
[38]	1995	10		5 years	100%	0%		100%	0%			
[38]	1995		90	5 years	7%	93%	?	?	27%			

Table 27 Postoperative results and types of preoperative deficits recorded in the literature

Reference	Year	No.	Anatomical type		Follow-up	Bladder			Motor/sensory		
			Filum	Conus		Improved	Stabilized	Worse	Improved	Stabilized	Worse
[78]	1976	31	31			3 of 3	0%	0%	9.1%	90.9%	0%
[196]	1985	24	14	10	Median 5 years	41.6%	58.4%	0%			
[126]	1986	50			Average 1 year	12%	?	?	40%	?	?
[97]	1988	21				52.3%	?	?			
[175]	1988	15				33.3%	?	?			
[94]	1990	42			Median 4 years	0%	?	?			
[164]	1993	8				37.5%	62.5%	0%			
[192]	1994	12	12			3 of 4	?	?	75%		

explained by the facts that our series included more patients than others, follow-up was much longer, and all patients have been systematically reviewed in the past 2 years preceding this study.

Why did nearly half the asymptomatic patients in our series who were operated on worsen with time? The answer is not clear. Retethering of the cord may be one reason, but we wonder whether it is the only one, or even the major one. It is striking to observe that postoperative long-term deterioration is a risk only in lipomas of the conus and meningomyeloceles, and never in spinal cord tumors, spinal cord arteriovenous malformations, or lipomas of the filum. All these operations nevertheless lead to similar postoperative scarring and adhesions. Why then, these different outcomes? The underlying myelodysplasia might play a part, and perhaps a major one. Myelodysplasia is indeed absent or minor in the case of tumor, arteriovenous malformation and even lipoma of the filum, but probably

constantly present at various degrees with meningomyeloceles and lipomas of the conus. From these observations, we suggest that risks of late postoperative decompensation might result partly from the degree of the underlying myelodysplasia. Postoperative adhesions might be only an additional contributing factor. In cases with minor myelodysplasia and severe retethering, recurrence of deficits could be improved or even cured by reoperation. Conversely, in cases with severe myelodysplasia reoperation might have little chance of improving patients' condition or preventing new postoperative deterioration.

Reoperations

Sixteen of our patients underwent reoperation (5.5%). Of these 16 patients, 6 were asymptomatic prior to first surgery. The first operation was uneventful in all cases. In 13 cases, postoperative worsening developed insidiously, and in 3 following a long period of apparent stabilization (3, 4, and 7 years). The average interval between the first and second operations was 6.5 years (range 1–7 years; median 6 years), similar to the intervals reported by others [77, 82, 96].

New or recurrent symptoms were motor deficits in the lower limbs in 4 cases, sphincter disturbances in 4 others, and both types of deficits in the remaining 8; 4 of these 8 patients also complained of diffuse and severe pain in the legs.

MR studies before reoperations showed a low-lying cord attached to the posterior aspect of the dura in all cases, a recurrent lipoma in 2 patients, a voluminous residual lipoma in 5, and a recurrent intralipomatous dermoid cyst in 1.

Reoperation was difficult in all cases owing to severe adhesions, as in the experience of Sakamoto et al. [163]. In 3 patients, surgery was stopped before complete freeing of the cord.

Postoperatively, 5 patients were clearly improved (31.2%) with follow-up of 1 year for 2, 2 years for 2, and 5 years for 1. In 7 cases (43.7%), the symptoms stopped evolving. There were 3 patients who continued to deteriorate (18.7%). Finally, 1 patient was worse after surgery (6.2%). Overall, patients who were symptomatic at first surgery did better than those who were asymptomatic. Of the latter, none improved after reoperation, 3 were stabilized with severe handicaps, 2 continued to deteriorate, and 1 worsened following the second operation. These 16 patients include 3 who were reoperated on twice because of continuing worsening of their symptoms, finally with no success.

In the literature, the rate of reoperation is similar to ours [45, 77, 163, 178]. Our results were grossly the same as those reported by Lhowe et al. [116], with 2 improvements and 1 failure, by McLone and Naidich [125], with respectively 33% and 14% improvement for the motor and sensory functions, and by Sakamoto et al. [163], with 1 improvement and 3 stabilizations. They were far from being as good as those achieved in the series of Hoffman et al. [79], Kanev et al. [93], and Herman et al. [77], all of whom report 100% improvement.

In summary, in our experience, reoperations, although efficient in some cases, often did not improve patients. Moreover, they were frequently technically difficult and not free of risk, not only in our experience, but also in that of Sakamoto et al. [163], who mentioned 3 cases of transient aggravation in 4 patients who underwent reoperation.

Indications for surgery

Surgical indications are a subject of controversy. Looking to the literature, however, there is consensus on the subject of symptomatic children and adults. For the children, all publications favor early surgery. In the case of adults, the few authors who have written on the subject are more reluctant, advocating surgery only when deficits are progressive [101] or even evolving severely [47]. Our opinion is slightly different. We believe surgery is mandatory at any age in the presence of deficits, even when these are nonprogressive. Our results showed that the majority of symptomatic patients, adults as well as children, benefited from surgery. In these patients, surgical risks are clearly counterbalanced by the severity of the disease. In adults, the proposal to wait for progressive symptoms before recommending surgery suggests that, after a certain age, the risk of deterioration is small or operations are ineffective, which is contradicted by our experience and that of many others [95, 109, 137, 141, 145, 172].

Discussions should only concern asymptomatic patients, because prophylactic treatments raise major medico-ethical problems. This is true for lipomas as it is for any other pathology. Examples of prophylactic treatments are rare in our medical practice and are justified only when three conditions are met: (1) low operative risks; (2) protective effect of the treatment; (3) severe disease.

To advocate routine surgery in asymptomatic lipomas, we therefore must be able to answer all these three basic questions: (1) Is surgery harmless? (2) Does surgery protect patients from further risk of deterioration? (3) Is the prognosis of lipomas poor? Answers to these questions must be considered separately for lipomas of the filum and lipomas of the conus.

Asymptomatic lipomas of the filum

In asymptomatic lipomas of the filum, the risks of surgery in all series including ours are almost nil. Almost all patients, as demonstrated in our study, remain symptom free,

and these good results are maintained over time. Does surgery really exert a preventive effect? Our data are insufficient for an answer. Nevertheless, we can postulate that surgery is at least harmless, and can reasonably put forward the hypothesis that the risks of surgery are smaller than those of the disease itself. For these reasons, routine surgery is acceptable and advisable and does not contravene the rules of ethics.

Asymptomatic lipomas of the conus

The policy for all patients in this series was to operate routinely on all asymptomatic lipomas of the conus, an attitude similar to that of the majority of neurosurgeons [7, 11, 12, 19, 32, 45, 47, 71, 75, 79, 84, 93, 94, 109, 111, 116, 117, 119, 126, 145, 153, 157, 175, 184, 185, 189, 193, 196]. However, our results have not been as expected, since approximately half the patients developed deficits despite surgery. Considering in addition the risks of postoperative deterioration – almost 4% in the case of lipomas of the conus – we cannot any longer advocate routine surgery in this group of patients; not only is treatment risky, but it also does not have much preventive effect. Perhaps in this series surgical risks would have been reduced by removing lipomas subtotally instead of radically as we always aimed. However, surgical treatment of complex malformations with severe adhesions or intralipomatous roots will always remain difficult and risky.

It is possible that the actuarial curve showing the slow postoperative deterioration with time in our patients (Fig. 37) is better than would have resulted from the spontaneous evolution of the disease. At present, a definitive answer cannot be given.

The natural history of disease has not been much studied. There is a vague feeling that aggravations are inevitable but, as already emphasized, the bias of recruitment in all series makes valid conclusions impossible, since healthy carriers of the malformation have never been taken into account. The only two reports on the spontaneous evolution of asymptomatic patients are contradictory. In the series of Bruce and Schut, 7 of the 8 patients deteriorated [32], while this was true of only 1 of the 10 in that of Lagae et al. [106]. The number of asymptomatic adult patients is also difficult to know. It could be high if one refers to both autopsy reports [17, 113, 124, 154] and radiological studies [31, 50, 131, 139, 187].

All these data show that surgery on asymptomatic lipomas of the conus does not fulfill the ethical rules prevailing in the other fields of medical practice.

Proposals and conclusions

At the outset of this study, we hoped to determine the frequency of lipomas of the filum and of the conus, to shed light on the natural history of the condition, to ascertain the short- and long-term results of surgical interventions, and to confirm the value of prophylactic surgery. As we feared, the project was too ambitious. We estimated the incidence of lipomas at 4–7.8/100,000 but this is probably an underestimate, as the number of asymptomatic patients who never seek medical attention is unknown. For the same reason, the true incidence of progressive deterioration cannot be established. We wish to stress the inherent bias in all previously reported series, as they all relate only to hospitalized patients. Despite these limitations the present study has a significant bearing on the management of spinal lipomas.

1. It emphasizes the hamartomatous nature of lipomas, which can vary in complexity. In particular, because these hamartomas may grow during infancy, regular MRI examinations are advocated during this period.

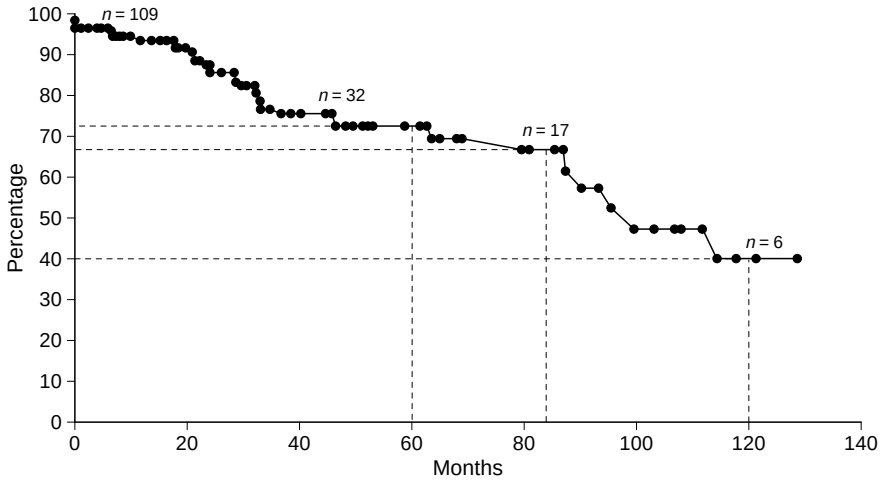


Fig. 37 Time to clinical deterioration after initial surgical intervention in patients with asymptomatic lipomas of the conus

2. It demonstrates that there are two distinct types of lipomas, a fact that is not sufficiently emphasized in the literature and has made interpretation of the results difficult. The first type is lipoma of the filum, for which surgery is not only beneficial in both the short and long term, but also has almost no morbidity. The second type is lipoma of the conus, which is much more complex, associated with more severe deficits, and for which surgery cannot be considered to be free of risk and is of questionable long-term efficacy.

3. A most important and surprising finding was that untethering of the spinal cord and debulking of lipomas of the conus fail to prevent the onset of late neurological deficits in almost half of the patients. This puts the rationale for prophylactic surgery in these patients in question. Are the results of surgery any better than the natural history of the disease? Yet again, the lack of basic information remains a stumbling block in the management of these patients. Until this question is answered, we are no longer able to advise prophylactic surgery in patients with conus lipomas. We now follow a policy of careful surveillance by a multidisciplinary medical team for these patients. At referral, they undergo EMG of the lower limbs and perineum, and urodynamic study. Infants are re-examined twice per year until the age of 2 and later once per year indefinitely. MRI will be repeated at the age of 6 months to ascertain that the lipoma has not grown in the interval. Urodynamic studies will also be repeated annually until the age of 3, or more frequently in the case of multiple urinary infections. Surgery for these patients depends upon one of four situations: (1) onset of deficits, foot malformation, amyotrophy, sphincter disturbances, abnormally brisk reflexes; (2) increase in volume of the lipoma; (3) abnormal urodynamic studies; (4) opposition of the family to this protocol of surveillance.

We await with interest the follow-up of this new cohort of unoperated patients. This follow-up will be analyzed in the next 2 years and statistically compared with the first cohort of operated patients. The new protocol will be stopped if the incidence of deterioration is significantly higher than that following prophylactic surgery. We hope that comparison of these two groups of patients will make it possible to formulate a scientific description of the natural history of the disease and to define the correct therapeutic attitude to asymptomatic lipomas of the conus.

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