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The transition from child to adult in neurosurgery

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With 8 Figures

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Abstract

The transition from child to adult is a growing concern in neurosurgery. Data documenting long-term follow-up are necessary to define this population's

healthcare needs. In order to evaluate the problems posed by the child-to-adult transition in neurosurgery, we have studied the neurological, functional and social outcome of patients treated in our department for tumor of the central nervous system, hydrocephalus or myelomeningocele, and followed beyond the age of eighteen years. A large number of patients suffered from chronic ailments, either sequelae of their initial disease, or delayed complications of their initial treatment, with significant morbidity. The mortality during adulthood was 4.6% in the tumor group, 1.1% in the hydrocephalus group, and zero in the spina bifida group. The proportion of patients employed in normal jobs was 35.6, 18.7 and 11.5% for tumors, hydrocephalus and myelomeningocele respectively. IQ score and performance at school generally overestimated the capacity for social integration. Based on these data and on the available literature, we tried to identify the problems and devise solutions for the management of the transition from child-to-adulthood transition. Many problems present during childhood persist to adulthood, some of which are made more acute because of a more competitive environment, the lack of structures and inadequate medical follow-up. The transition from child to adult must be managed jointly by pediatric and adult neurosurgeons. More clinical research is required in order to precisely evaluate the problems posed by adult patients treated during childhood for the different neurosurgical diseases. Based on these data, a concerted trans-disciplinary approach is necessary, tailored to the specific needs of patients suffering from different diseases.

Keywords: Myelomeningocele; cerebral neoplasms; hydrocephalus; outcome; age.

Introduction

With recent advances in pediatric neurosurgery and the resulting increase in survival, the care of adult patients treated for pediatric neurosurgical disease during childhood has become a new field of activity. In many of these patients, although the initial disease is cured, or at least well controlled by various treatments, several problems are present in adulthood. In some patients, these problems become worse with advancing age because of premature degenerative ailments, e.g. patients with spina bifida loosing ambulation. In other diseases, like hydrocephalus, the initial problem is only temporarily settled by a prosthetic device (the valve) or a palliative procedure (endoscopic third ventriculostomy), both of which may present with delayed failure. Finally new problems may arise as a delayed consequence of the initial treatments, such as brain lesions after irradiation for brain tumors, or as complications of initial treatment, like meningeal infection, or because of an inborn defect predisposing to disease, like the phakomatoses.

Adult patients with pediatric neurosurgical antecedents pose difficult management problems: their medical history is often complicated, and may be

difficult to reconstruct because of missing records. Another problem is that neurosurgical subspecialties tend to separate from each other, and in particular, pediatric neurosurgery tends to become the exclusive field of pediatric neurosurgeons. As a result, many adult neurosurgeons do not have any longer the necessary experience to treat specific diseases, e.g. the dysraphisms. The management of the child-to-adult transition poses thus several problems regarding medical competence, training, and availability of care for these patients.

Recently, the American academy for pediatrics has issued recommendation for the transition from child to adult, to be implemented and adapted to the different fields of child-oriented care [1]. Neurologists have begun to show concern over this problem [35]; however, we were unable to find any neurosurgical literature dealing with this subject.

By focusing on adult patients treated during childhood for three categories of diseases – myelomeningocele (MM), tumors, and hydrocephalus – we have tried to evaluate the magnitude of the problem and explore some possible solutions.

Example of three groups of patients

We selected the cases of patients with tumors of the central nervous system, myelomeningocele and hydrocephalus (the latter group overlapping widely with the other two) treated in our department, who had reached the age of eighteen. We chose to study the adult outcome of these three groups because they represent large shares of the accrual of pediatric neurosurgical departments, and their medical follow-up is prolonged into adulthood because of specific problems.

In our institution, we are in a situation of virtual monopoly for pediatric neurosurgical diseases for a four-million population, and we have the opportunity to follow these patients beyond their entry into adulthood. Medical data have been stored in a quasi-prospective fashion for more than three decades; our medical database includes now over 5,000 patients with pediatric neurosurgical diseases, of which over 800 were followed beyond child age.

We evaluated the overall functional outcome using the semi-quantitative Glasgow Outcome Score for hydrocephalus and MM patients, and with the Karnofsky independence scale for tumor cases; more specific ailments (like endocrine disorders) were rated in a binary fashion (present or absent). The social outcome of the patients having completed their training was rated as "normal employment" (which includes child-rearing for mothers-at-home); "sheltered employment" (when the job was obtained by legal protection for the handicapped); "occupational activity" (when the job was part of a therapeutic

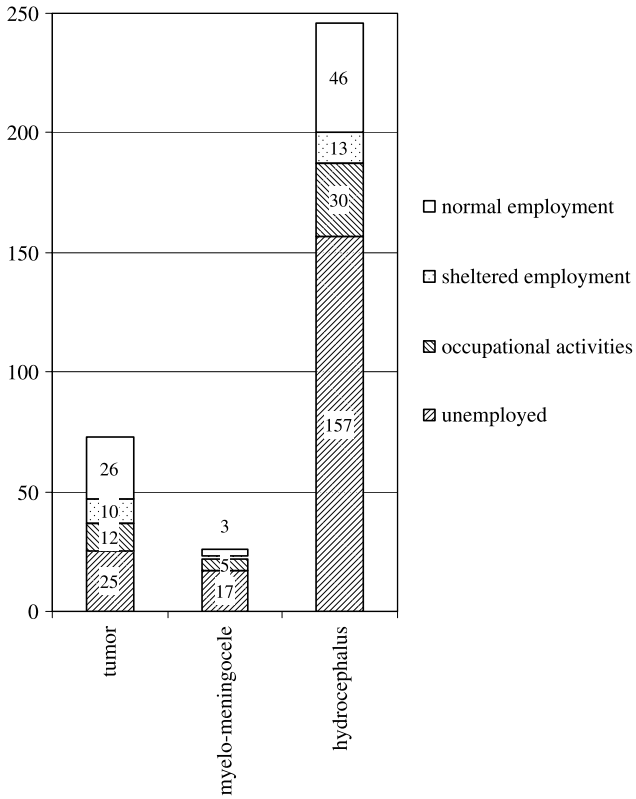

Fig. 1. Diagram showing the proportion of patients normally employed, employed in a sheltered environment, having occupational activities, and unemployed, in the three diseases studied. The particularly low employment rate among patients with myelomeningocele can be explained by the disturbance of schooling due to repeated hospitalizations during childhood, in addition to their neurological handicap

program rather than market-driven); and unemployed. The social outcome in the three groups of patients studied is summarized in Fig. 1.

Tumors

Personal series

From our series of 1065 children treated for tumors of the central nervous system since the advent of CT scanner, we selected 213 cases followed beyond the age of 18; 277 patients had died before reaching that age, and the rest are either still of child age, or lost to follow-up. For the purpose of this study, tumors were regrouped: cerebellar astrocytoma (34); lobar low-grade glioma

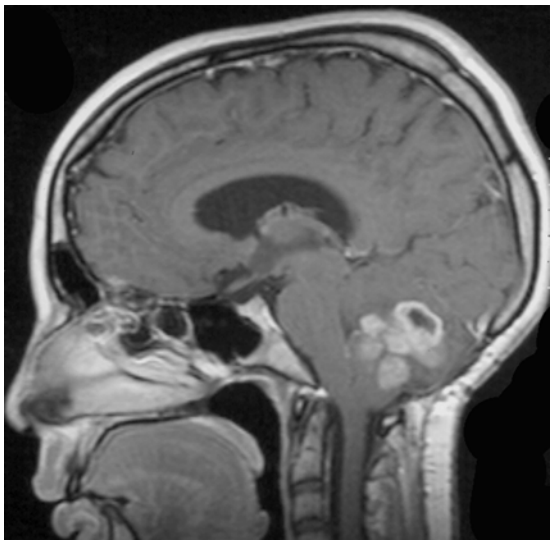

Fig. 2. 21-year old female, treated age 18 months for medulloblastoma with total removal and external irradiation (45 Gy); she developed a cerebellar high-grade astrocytoma, which was excised and treated with chemotherapy, but recurred after five months, and was the cause of demise 10 months after surgery

(22); optic pathway glioma (21); diencephalic low-grade glioma (19); brainstem low-grade glioma (18); malignant glioma (10); medulloblastoma (34); ependymoma (19); craniopharyngioma (14); pineal tumor (13); subependymal giant-cell astrocytoma (6); and schwannomas (3). The mean age at diagnosis was 10.0 years, and the mean duration of follow-up was 134 months.

Twelve patients (5.6%) died after having reached adult age: 9 because of tumor progression, one after surgery for recurrent craniopharyngioma, one because of a radio-induced malignant glioma (Fig. 2), and one because of radiation-induced vasculitis (Fig. 3). The overall clinical outcome at the last visit was evaluated with the Karnofsky independence score: 75 patients (35.2%) had a score of 100 (asymptomatic), 33 (15.5%) had a score of 90 (some symptoms but normal activity), 36 (16.4%) had a score of 80 (some symptoms but able to go to work or school), 20 (9.4%) had a score of 70 (independent at home but no outdoor activity), and 12 (5.6%) had KNK scores between 60 and 10 (diverse degrees of dependence on a third person and altered health status). In Fig. 4, we report the number of patients having behavioral, cognitive, memory, motor, endocrine, sensory deficit, as well as the presence or absence of obesity, epilepsy, spinal or shunt problem; those patients who had none of these afore-mentioned ailments were rated as having "no problem": only 37 patients (17.4%) were thus completely normal. The discrepancy between 37 "completely normal" patients and 75 patients with a Karnofsky score of

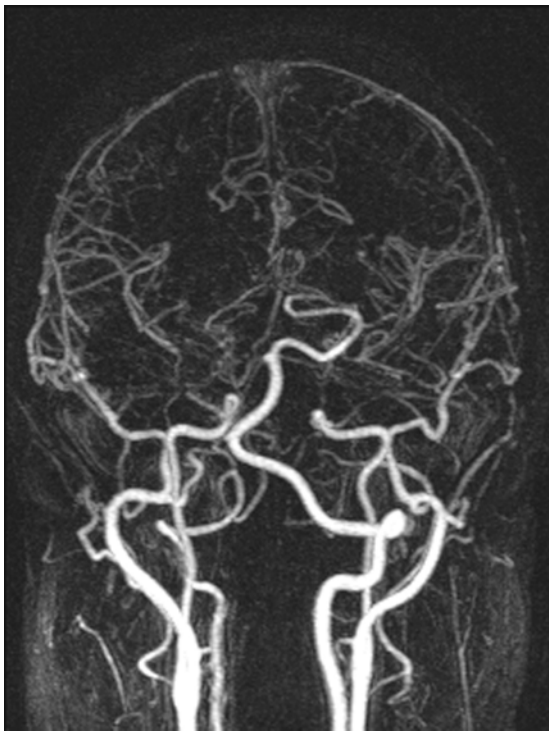

Fig. 3. 23-year old male, treated age 11 months for optic glioma, with subtotal resection followed by irradiation at the age of 18 months. He has severe visual loss and pursues occupational activities (upholstery). His MRI shows severe post-irradiation angiopathy with complete occlusion of the left posterior cerebral and both internal carotid arteries

100 was explained by patients having mild clinical signs (like ataxia) but no awareness of it, or patients with epilepsy well controlled under medication. Figure 5 summarizes the number of patients in each group of tumors, with the number of patients having "no problem" in each group: totally asymptomatic patients were found mostly in the cerebellar astrocytoma and low-grade lobar glioma groups.

Formal IQ testing was performed in 76 patients: the mean full-IQ score was 81.9, and 46 patients (59.7%) had full-IQ scores at or above 80. At the time of evaluation, 34% of the patients had completed high school, and 13% were or had been in university. Among 73 patients having completed their training, 36% were employed on a normal job, 14% were employed in a sheltered environment, 16% had occupational activities, and 34% were unemployed (the present official unemployment rate in the normal population is around 9% in our country).

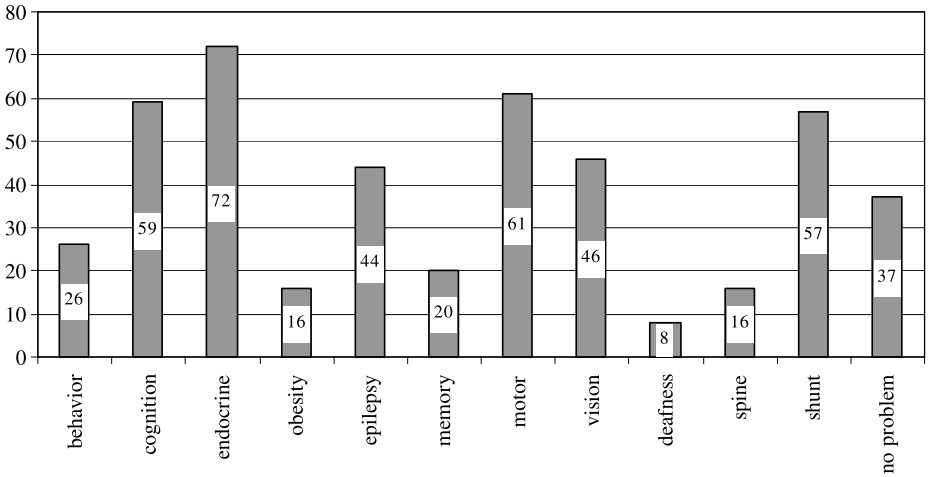

Fig. 4. Prevalence of the different sequelae in adult survivors of childhood tumors of the nervous system. The patients having ''no problem'' were the group of patients having none of the different ailments detailed here

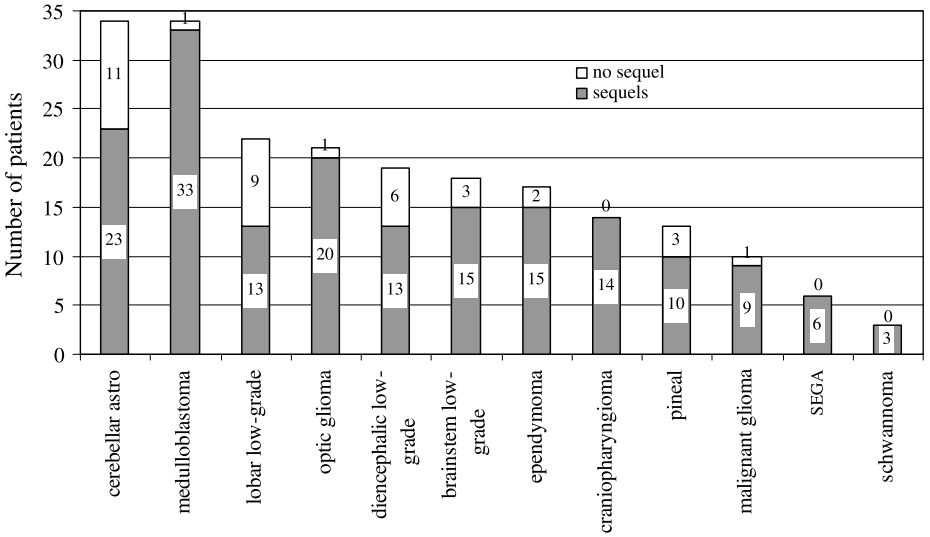

Fig. 5. Presentation of the different groups of tumors, with the proportion of patients having or having not neurological sequels. As expected, medulloblastoma, optic pathway glioma and craniopharyngioma were associated with the highest morbidity. However, contrary to common wisdom, cerebellar astrocytomas and low-grade lobar gliomas left sequels in more than half of cases, although generally not severe enough to impede normal life. (*SEGA* subependymal giant-cell astrocytoma)

Survival and oncological outcome

The long-term outlook of patients treated during childhood for brain tumor is characterized by an « appreciable burden of morbidity », with higher morbidity and poorer quality of life, compared with other childhood malignancies [10]. Nicholson calculated that mortality during the fourth decade of life was increased four-fold for patients with a history of brain tumor during childhood [24]. Although late tumor progression is the most likely cause of death, other causes, mostly treatment-related, are to be expected. Second tumors can be radio-induced, or due to a genetic predisposition (phakomatosis). In our experience, among 419 children irradiated for brain tumor, 14 developed tumors or cavernomas, and survival analysis showed that the prevalence of radio-induced tumors was 4.2% after ten years (unpublished data); this means that many of these tumors occurred in adults. The risk of radio-induced tumors could be higher when patients are irradiated at a younger age, because of a higher number of pluripotent cells [21]. Some diseases appear to predispose patients to develop radio-induced lesions, like neurofibromatosis type 1 [6] and Gorlin disease [2].

Functional outcome

Our data show that even among fully functional patients, many are not asymptomatic. The chief causes of this morbidity are the tumor itself, surgery, and irradiation. The most prominent sequels are neurological, neuropsychological, endocrine and sensory disturbances [25]; however, as shown in our Fig. 4, every facet of the functions of the nervous system can be more or less severely affected.

Neuropsychological sequels vary in severity according to tumor location, patient's age, and irradiation [23]. These results can be ascribed to progressive lesions of the central nervous system, as well as to difficulty increasing with age, as the school environment becomes more demanding. Hope-Hirsch and co-workers have alerted about the occurrence of progressive loss of IQ after irradiation for medulloblastoma [17]. Palmer calculated that the mean loss of full-scale IQ adjusted for age was 2.55 point per year in patients irradiated for medulloblastoma; the effect was especially marked for children who were younger at the time of irradiation, and for those who received higher irradiation doses [26]. This prolonged and sustained decline in intellectual function implies that a prolonged follow-up is necessary to evaluate its full extent [20]. Cognitive deficits are also related to the location of the tumor, and to the cerebral damage caused by the tumor and its resection. Recently, the role of the cerebellum in cognitive functions has been highlighted; Grill has shown that a lower performance IQ in medulloblastomas was associated with splitting of the vermis and damage to the dentate nuclei of the cerebellum, whereas lower verbal IQ was

associated with hydrocephalus [13]. Patients with supratentorial tumors often have dysfunctions of executive tasks like programming strategy and inhibition, amounting to a « deficit in social competence » [24]. IQ tests show their limits in predicting the patient's adult outlook, because these problems, which can represent a major handicap for adult life, are most difficult to assess [3, 20]. Long-term data on adult independence score and social achievement are rare in the literature. Our series show a wide gap between independence score (67% were autonomous for outdoor activities) and actual academic and social achievements (only 36% were normally employed). This can perhaps be explained in part by repeated hospital stays disturbing schooling, but also, more probably, by more subtle sequels of higher brain functions, which may constitute a major handicap in a competitive environment. The gap might be even wider if we could compare the patient's achievements to what could be expected from his familial background. Our dataset does not give access to such information, and more detailed studies would be necessary to precise this point.

Endocrine problems are another major group of complications in brain tumors, resulting from damage to the hypothalamic-pituitary axis caused by the tumor, surgery and irradiation [34]. After having complicated the patients' growth and pubertal development, hypopituitarism continues to represent a sizeable problem during adulthood. Growth hormone deficiency is almost universal after brain irradiation of more than 30 Gys [11], and puts patients at higher risk of osteoporosis, muscle wasting, obesity, cardiovascular and cerebrovascular diseases. The benefits of treatment with growth hormone during adulthood on health risk factors and general fitness have been demonstrated [16], however the financial cost as well as lack of motivation for a lifelong injected treatment have limited its spread. Obesity is associated with hypothalamic damage due to the tumor, surgery, or irradiation. It is considered to result

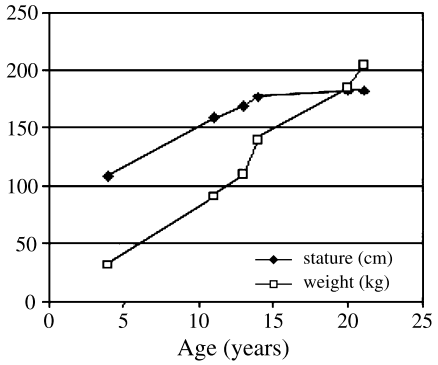

Fig. 6. Eighteen-year female treated since the age of four for craniopharyngioma, with surgery alone. At the time of the fourth tumor recurrence, her weight was 204 kg and rising

from lesions of the ventromedial nuclei, which mediate the blood-borne information from insulin, ghrelin and leptin [11]. The result of these lesions can be compounded with growth hormone deficiency and obesity-inducing treatments (like steroids or Valproate®) to produce morbid obesity. In craniopharyngiomas in particular, all these factors concur to make obesity a major concern, affecting up to 50% of patients [33]. We experienced a case of craniopharyngioma who reached a peak weight of 204 kg (450 lb), aged eighteen, at the time of her fourth recurrence (Fig. 6). Strategies seeking to avoid damage to the hypothalamus should be developed, and strict measures to contain weight gain should be enforced early during childhood, in order to avoid such nightmarish developments. Fertility problems are also common in adults treated for brain tumor during childhood, resulting from hypothalamic damage as well as gonadic toxicity due to chemotherapy [11]. Our series confirm a low fertility rate, only 14 of our 174 adult female patients (8.0%) having given birth to 16 children. Other possible obstacles to reproduction are teratogenic drugs (chief among them being antiepileptic drugs), genetic disorders like phakomatosis, and obstacles to delivery due to pelvic deformity or neurological deficits. However, the prevalence of inborn defects in offspring of patients treated for cancer during childhood (3.3%) does not appear different from that of the general population [12].

Implications for initial treatment of the tumor

The price tag of survival in brain tumor patients may look prohibitive. The elaboration of oncological protocols should include the evaluation of long-term morbidity [20]. The cause of long-term morbidity in tumor patients can generally be ascribed to one of the "three villains": the tumor, surgery, and irradiation. Radiotherapy is especially deleterious in infants, as well as patients predisposed to radio-induced lesions, like NF1 and Gorlin disease, and should thus be considered a last resort in these patients. Early recognition of morbidity related to radiotherapy and surgery has led to the development of alternative treatments with chemotherapy, for example for infiltrative low-grade astrocytomas (BBSFOP protocol) and germ-cell tumors. The quest for lower morbidity should not let forget, however, the first aim of treatment, which is the eradication (or at least stabilization) of the disease. In the long run, the risk of tumor recurrence, and the risks associated with the treatment for recurrent tumor, should be taken in account. For example, radical resection for craniopharyngiomas is known to be associated with a risk of severe morbidity, and subtotal resection followed by irradiation have been advocated [30]. Conversely, surgery for recurrent craniopharyngioma is technically demanding and risky, especially after irradiation, and our data show that operative morbidity was highest in case of surgery for tumor recurrence; this would incite to perform maximal initial resection whenever it can be done safely [8]. This question

remains open, until more long-term data are available. In other tumors, like ependymomas, aggressive resection is recognized as the main factor influencing outcome [37]; in some cases however, resection cannot be carried out without unacceptable morbidity, and we have to rely on postoperative treatments. Because the decision to pursue or not complete resection has to be taken on-the-spot during surgery, it is important that we try to settle these questions in advance, based on long-term oncological and functional results.

Myelomeningocele

Personal series

Among 452 patients with MM treated in our department, we selected 38 patients (19 male and 19 female) aged between 21 and 23 years, and surveyed them with a detailed questionnaire and interview by phone call. The quality of life was studied using the SF-36 health survey, which is a validated scale based on a questionnaire regarding physical health, fitness, pain, mental health, social skills, and emotions [40], and has been translated and validated in French language [28].

Thirty-three patients (86.8%) had a CSF shunt. The average number of reoperations for shunt failure was 2.5 per shunted patient; 6 patients had undergone surgery for Chiari malformation, 14 patients (36.4%) for tethered spinal cord, and 5 (13.2%) for scoliosis. Overall, the average number of surgeries (including initial closure and shunt) was 10.2 per patient (1 to 28).

At last control, 14 patients were able to walk for more than 500 meters, four were able to walk between 50 and 500 meters, 9 were able to walk for less than 50 meters, and 11 were non-walking. Among 31 patients initially using walk as their regular mode of ambulation, 8 had lost walking between the age of 7 and 20 years (Fig. 7). Although loss of ambulation was often multifactorial, the chief cause was considered to be prolonged immobilization after orthopedic problems in four cases; obesity, spinal cord tethering, and degradation due to Chiari malformation, and accidental brain damage after drowning in one case each.

Among 16 patients who underwent formal neuropsychological testing, mean verbal IQ was 96.1, performance IQ was 85.8, and global IQ was 92.1. Eleven patients had no school degree, 5 had completed primary school, 14 were in high school, and 8 were at university. At last control, 12 patients were studying, 3 were working on a normal employment, one in a sheltered environment, 5 had occupational activities, and 17 had no outdoor activities. The quality of life studied using the SF-36 health survey showed that although the health and vitality indices were as expected lower than in the controls; however, surprisingly, the indices relating to psychological and emotional domains scored on average better than in the control population published by Pergener [14].

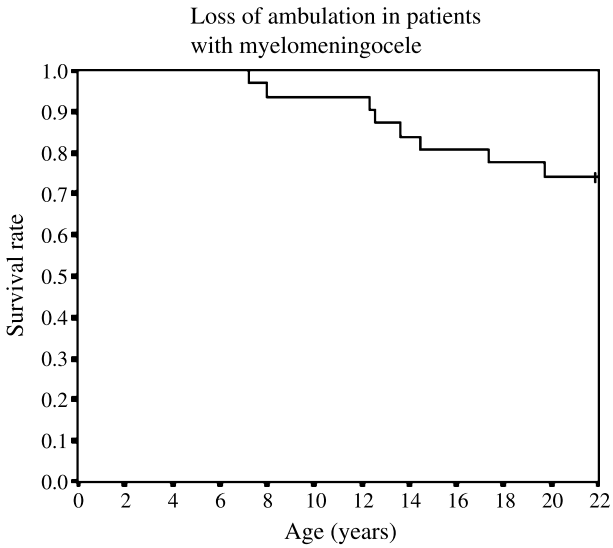

Fig. 7. Diagram showing the incidence of loss of ambulation among 36 patients with myelomeningocele surveyed: among 31 initially ambulating patients, 8 lost walking as their regular mode of ambulation between the age of 7 and 20 years. The 20-years actuarial survival rate was 74%

Discussion

Because of antenatal diagnosis and the common practice of termination of pregnancy, MM is a disappearing disease in pediatric neurosurgery; the bulk of patients with MM are now slowly but surely becoming adults, and their needs must be addressed. Adult patients with spina bifida are considered "the most neglected individuals in the population with neurosurgical disease" [22]. The population followed in the long term is biased compared to the initial population, because the most severe cases died before reaching adulthood, and the less severe, often not shunted, tend to abandon medical follow-up [18]. Even during adulthood, patients with MM have an excess mortality, mostly because of shunt failure [4].

Although the initial motor deficit is a direct function of the anatomical level of the MM, children who have managed to acquire walking can lose it lately because of tethered cord, syringomyelia, or neuro-orthopedic problems. Bowman considered that loss of ambulation occurred mostly during childhood and that "patients who remain mobile in their teens continue to ambulate . . . in their young adult years [4]. Our data do not concur, as shown in our Fig. 7. We have some reasons to fear that with increasing obesity and neuro-orthopedic problems, compounded with premature skeletal aging, loss of ambulation will become more and more prevalent in this group of patients.

Tethered cord is a delayed complication of MM occurring mostly during late childhood and adolescence [32]. However four patients in our experience underwent spinal cord untethering between 24 and 37 years. The rarity of this condition in adults may be due to the lack of information amid adult neurosurgeons, and the number of adult cases is likely to grow in the future. In order to be performed safely, spinal cord untethering requires careful indications, surgical skill and experience, and we think that pediatric neurosurgeons should continue to operate adult patients with tethered spinal cord.

Academic and social achievements are low in the MM population for several reasons. Their IQ is frequently low [27], especially performance IQ [15]. Lower IQ may be due to associated brain malformation, and/or hydrocephalus, with repeated episodes of raised intracranial pressure [18]; however the relation between the number of shunt revisions and social achievements has not been confirmed [15]. The correlation of hydrocephalus with schooling and social achievement in MM patients is blurred because patients with low-level MM are often doubly blessed with shunt-independence as well as autonomous walking. Even with a normal IQ, schooling may be difficult: our data show that the vast majority of patients underwent multiple surgical procedures during childhood. In addition, because of their motor deficiencies, these patients often had to attend schooling in rehabilitation centers. In itself, this hectic curriculum may be responsible for low achievements. Also, a sense of discouragement and lack of motivation when reaching adulthood often appears to be a potent obstacle to these patients' social life.

These data on MM are important for antenatal counseling, because the decision to continue or interrupt pregnancy must be assisted by medical evidence. An illustrative case is the only male MM patient in our experience who was able-bodied enough to sire a child; when it turned out that the fetus had spina bifida too, his decision to have the pregnancy terminated was immediate and final. On the other hand, our study found that SF-36 health survey indices relating to psychological and emotional domains were at least as high in MM patients as in healthy controls; this surprising result shows that in spite of their handicaps, a degree of well-being is undeniable in these patients.

Hydrocephalus

Personal series

We selected 450 patients shunted for hydrocephalus during childhood, and aged more than 18 years at last control. The median age at shunt insertion was 8 months. Overall 1188 shunt revisions were necessary, the mean number of shunt revisions being 2.6 per patient; only 65 patients (14.4%) had no shunt

revision. In 15 patients, the first shunt revision was performed more than 20 years after insertion. Eighty-two episodes of shunt infection occurred in 70 patients; the rate of infection was thus 15.6% per patient and 5.0% per operation.

Ten patients died between 18 and 34 years: 4 of tumor progression, 2 of medical causes related to the cause of hydrocephalus, one of radio-induced lesions, one because of shunt infection, one of ascertained shunt failure, and one of unexplained sudden death (possibly caused by shunt failure). Five patients (1.1%) can thus be considered to have died of hydrocephalus-related causes. Overall evaluation following the GOS showed that 184 patients (40.9%) had normal activity, 70 (15.6%) had a mild handicap, 141 (31.3%) had a more severe handicap, and 5 (1.1%) were vegetative. Schooling had been; normal in 149 cases (33.1%); difficult in 70 (15.6%); delayed in 29 (6.4%); special schooling for the handicapped in 121 (26.9%); and no schooling had been possible in 32 (7.1%). At last control, among 246 patients having completed or abandoned school, 46 (18.7%) were working on a normal job, 13 (5.3%) were working in a sheltered environment, 58 (23.6%) had occupational activities, and 157 (52.4%) were unemployed or unable to have outdoor activities.

In a previous study [39], we determined in a binary logistic regression analysis which factors influenced independently the schooling and overall outcome of shunted patients. The results are shown on Table 1. Most of these factors pertained to the cause of hydrocephalus (post-hemorrhagic, post-meningitis, or due to spina bifida), but shunt infection was also a major independent factor influencing both schooling and overall outcome. These results highlight the fact that complications of treatment play a major role on adult outcome.

Table 1. *Determinants of schooling and overall outcome in hydrocephalus: binary logistic regression analysis [39]*

Variables in the model	Outcome (GOS = 1 or more)*	Schooling (normal or not)†
Post-meningitic hydrocephalus	<0.001	<0.001
Post-hemorrhagic hydrocephalus	0.004	NS
Myelomeningocele	<0.001	<0.001
Prematurity	NS	NS
Antenatal diagnosis	NS	NS
Shunt infection	0.002	0.009

* *and* † Significance of the model: $p < 0.001$. *GOS* Glasgow Outcome Score; *NS* not statistically significant. These data show that the prognosis is mostly dependent on the cause of hydrocephalus, but also on complications of treatment like shunt infection.

Discussion

Shunt outcome

The risk of shunt obstruction is the main burden for shunted patients who otherwise do well. It may be the cause of sudden death, with a risk estimated as high as 1% a year [29]. Repeated episodes of shunt obstruction have also been deemed responsible for poor functional outcome [18]. We consider that the potentially devastating complications of shunt obstruction make a regular and life-long follow-up necessary for all hydrocephalus patients; whenever an asymptomatic shunt failure is detected, we perform elective shunt revision, unless the patient can be determined as shunt-independent [36]. Conversely, symptomatic shunt failure is proof that the patient is shunt-dependent. When a shunted patient has never presented with shunt failure, the question whether the shunt might have been blocked long ago but the patient has become shunt-independent may be legitimately raised. The actuarial survival curve after shunt insertion shows typically a binary curve, with early failure due to surgical causes, and delayed failure due to interactions between the shunt and the patient. However, close examination of the actuarial survival curve prolonged beyond 20 years

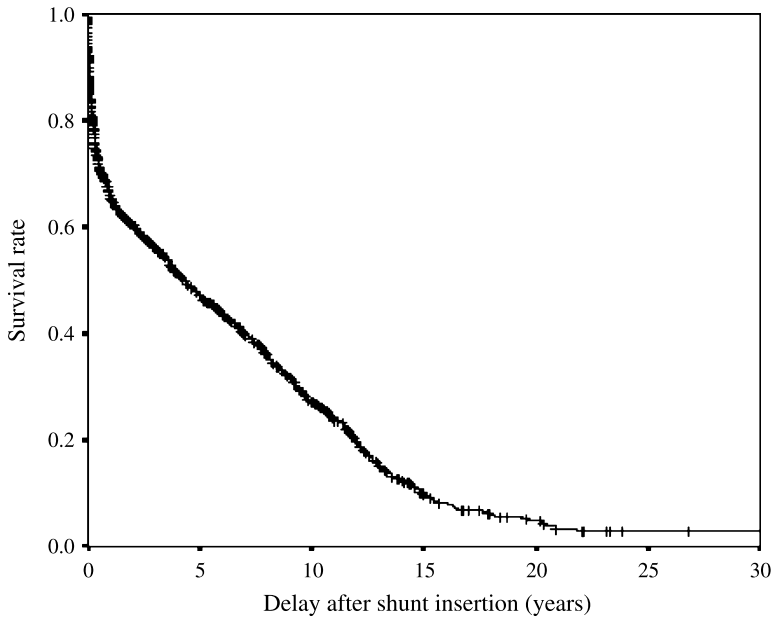

Fig. 8. Event-free survival after first shunt insertion: the curve shows that, although most cases of shunt obstruction occurred within the years following shunt insertion, the first shunt failure could occur after more than 20 years. Although very long follow-up introduces a selection bias, this diagram indicates that a long period of time without shunt failure cannot be equated with shunt independence

shows that shunt failure can occur very late (Fig. 8). In our experience, 15 patients had their first shunt revision performed more than 20 years after insertion. This shows that shunt-independence cannot be guessed from the absence of shunt failure, but has to be proved by a controlled procedure. Our routine procedure includes a shuntogram followed by shunt ligature, then shunt removal a month later if the ligature is well tolerated [36]. Regarding endoscopic third ventriculostomy (ETV), the risk of delayed obstruction appears more and more to pose problems similar to shunts, with catastrophic consequences in some cases; close radiological and clinical follow-up is warranted after ETV as well [9].

Although shunt infection is generally considered to result from intra-operative contamination, we found in a previous study that 12% of episodes of shunt infection occurred more than a year, and up to 15 years after previous surgery [39]. The causes of delayed shunt infection include hematogenous seeding, spontaneous bowel perforation, abdominal surgery, accidental penetrating trauma, but also possibly the prolonged persistence of dormant bacteria. We calculated that the incidence of spontaneous bowel perforation was 0.1% a year; we also found that it occurred more often in patients having walking difficulties, like spina bifida, which suggests that it might be facilitated by abnormal bowel motility [38].

Intellectual outcome

The impact of shunt obstruction on IQ has been suggested by Hunt, who noted that patients with more shunt revision had poorer achievements than patients with less shunt revisions, or without shunt; however the statistics backing this assertion were poor [18]. Most authors consider that the intellectual outcome depends on the cause of hydrocephalus rather than on the number of shunt revisions and the occurrence of shunt infection [5]. In particular, brain malformations, neonatal meningitis or ventricular hemorrhage have a major impact on development [29, 31]. Our opinion is that the number of shunt revisions does not necessarily indicate a high number of episodes of raised intracranial pressure, but may instead reflect the care with which patients are followed. A good illustration of this is provided in the series of patients with MM from Chicago, who underwent a high average number of shunt revisions (4.3 per patient), because shunt obstruction was systematically considered as a cause for neurological deterioration in these patients [4]. Shunt infection is classically associated with loss of IQ [19], although this view has been challenged by Casey [5]; in our series, we found that shunt infection had a major and independent impact on both schooling and quality of life of shunted patients [39]. Overall, we consider that IQ measurements overestimate the school abilities, which in turn overestimate the patients' professional achievement. Although Sgouros reported that 56% of shunted patients are normally

employed [31], our results are much inferior, which might reflect differences in patient accrual or in toughness of the labor market.

Organization of the transition from child to adult in neurosurgery

In 2002, the American Academy of Pediatrics stated that « by the year 2010 all physicians who provide primary or subspecialty care to young people with special health care needs 1) understand the rationale for transition from child-oriented to adult-oriented health care; 2) have the knowledge and skills to facilitate that process; and 3) know if, how, and when transfer of care is indicated » [1]. These recommendations apply to all fields of child-oriented care, including pediatric neurosurgery; up to now, the literature dealing with the transition from child to adult in neurosurgery is conspicuously lacking. The present report aims at pointing out some of the problems regarding the necessary "knowledge and skills", and raising awareness of our responsibility in ensuring the transition from child to adult. In European healthcare systems, the financial aspect of the child-to-adult transition is not as large an issue as it may be in North America; with the current trend toward liberalization in our healthcare systems however, we must be wary that the rights of our patients for continued care are respected.

Relation of pediatric to adult neurosurgery

The treatment of many neurosurgical diseases often transforms an acute and deadly disease into a chronic, lifelong condition. Tuffrey stated that « diseases of childhood are becoming diseases that begin in childhood and continue into adult life » [35]. As is the case for many chronic diseases, the transition from child to adult has become a major problem in neurosurgery. In her paper on long-term follow-up of spina bifida, R. Bowman stated that "one of the greatest challenges in medicine today is to establish a network of care for these adults with spina bifida" [4]. Although the same can be said of many other pediatric diseases, we neurosurgeons must be part this endeavor.

In the majority of neurosurgical centers in France, neurosurgeons are not age-specialized. A commonly accepted view in neurosurgery is that, apart from a few specifically pediatric fields (craniofacial, dysraphism, and neurosurgery in infants and newborns), children are not much different from adults. We do not share that view for several reasons. First, many diseases which look similar behave completely differently in children and in adults (e.g. subdural hematomas), while others, which have marginal importance in adults are major issues in children (e.g. arachnoid cysts). Second, the clinical presentation in non-verbal or unreliable patients requires specific communication skills for the diagnosis and medical management. Third, the small body size and immaturity

of tissues implies specific surgical and anesthetic techniques. Fourth, the patient's process of development and learning implies protracted follow-up and evaluation of long-term consequences. The downside of developing pediatric neurosurgery as an autonomous specialty is that it may easily lose contact with rapidly evolving fields (like spine or vascular surgery). Another problem is that pediatric neurosurgeons may become unable (because of limitations of resources or tight regulations), or unwilling (because of a busy schedule), to take care of patients becoming adults. The worst solution would be to simply discharge the patient to adult colleagues. Several halfway solutions have emerged in different places. In non-specialized centers, generally one adult neurosurgeon in the team is designated to take care of most pediatric cases (especially urgent cases), while more specifically pediatric (and non-urgent) cases are referred to another, more specialized centre. In specialized centers (like ours), the segregation is not complete, pediatric neurosurgeons having a "double citizenship" which allows them to continue to take care of their patients beyond their entry into adulthood, ensuring a relatively smooth transition.

Another possibility would be to subspecialize by disease instead of by age: this is already the case in some places for epilepsy surgery and for surgery of movement disorders; one can imagine that this concept could be extended to other fields, like the skull-base, spine, cerebrovascular, trauma, oncology, hydrocephalus . . . . However, a single patient's disease often encroaches on several subspecialties; CSF problems in particular are almost ubiquitous in their association with any other disease in pediatric neurosurgery. In addition, the problem of follow-up of the patient's development would require some form of centralization by a truly pediatric neurosurgeon, or a pediatrician dedicated to neurosurgery. The main obstacle to such an organization would be a shortage of manpower, because devoting one neurosurgeon (or ideally a team) to all the potential neurosurgical subspecialties would be beyond the reach of even in the largest centers.

Offer/demand of care

The offer of care for patients becoming adults is often insufficient. As mentioned above, adult neurosurgeons do not always have the necessary expertise for highly specific diseases, and pediatric neurosurgeons are not always available for adult follow-up. A major problem is the lack of adult structures for the care, professional training and employment of adults with handicaps. Grownup patients and their families are often dismayed when reaching the adult age limit, facing a medical and institutional vacuum they could hardly imagine while in the (relative) abundance and coziness of childhood care.

On the other hand, the demand for care from these patients is not always clear. The sense of discouragement often resulting from years of struggle against handicap often leads patients to abandon medical follow-up. In some cases,

these patients come to us for motives like recognition of a handicapped status, or demands for social benefits, which we can hardly satisfy, and are not aware of possibilities of medical treatments that we can offer. This mismatch between demand and offer of care needs to be clarified by exchange of information in both directions between patients, notably through their associations, and physicians.

Networking

From the present state of neurosurgery in our country, we are convinced that progress can be made through the training of young neurosurgeons, who should devote some part of their curriculum to pediatric neurosurgery, and dialogue between pediatric and adult neurosurgeons for example during post-graduate sessions. We need to spare some time for this necessary dialogue, during which adult neurosurgeons, pediatric neurosurgeons, and above all the patients, have much to gain. Also, if we are to make some progress in this field, more clinical data should be gathered and published; some of the above-mentioned examples show that the transition from child to adult is a rich field for useful clinical research.

The transition from child to adult can be at least as chaotic in specialties neighboring neurosurgery. For example, neuropediatricians stem from a general pediatric training, whereas neurologists generally do not have any training in pediatrics. The same can be said for intensivists, oncologists, and physical therapists. Bridges between the pediatric and adults side of these many specialties often remain to be built. Neurosurgeons caring for pediatric patients becoming adult have to create their own network, and establish preferred relations with colleagues in these neighboring specialties. In so doing, neurosurgeons play, in its most valuable sense, the role of a general practitioner. Establishing and motivating such a network can be both arduous and rewarding.

Conclusion

The transition from child to adult is a crucial period of everybody's life; social and professional integration requires the best from an individual's highest intellectual functions, as well as physical fitness. Patients with a history of neurosurgical disease during childhood often have cognitive, sensory or endocrine sequels that make them especially vulnerable and result in inferior social and professional achievements.

The fact that so few of our patients become successful adults in a competitive environment may be somewhat sobering. Like many physicians dealing with children, we indulge in self-complacency, in the belief that the children we have saved will become fit and productive adults, and that our actions are profitable for society. Clearly, if healthcare planners caring only for profitability

were to learn from these data, their reaction would be to cut down on the funding of pediatric neurosurgery. It should be stated loud and clear that the benefits of care for children as well as for handicapped adults should not be counted in economic terms. Its financing should thus be written in the losses column of the balance sheet; in the profit column however stands social equity and solidarity, the value of which is incalculable. How large a part of its gross domestic product our society is ready to spend on solidarity may be a vital choice for the future.

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