M. Memet Özek • Giuseppe Cinalli • Wirginia J. Maixner

# The Spina Bifida
## Management and Outcome

**Forewords by**
C. Sainte-Rose
C. di Rocco

**Preface by**
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Springer

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*For 700 little candles having shed light on my path and assisted me to do it right*

M.M. Özek

*To my mother, to Fabrizia, Francesco and Maria Allegra*

G. Cinalli

*To my parents, who through their love have allowed me to realise my dreams*
*To my daughter, the most beautiful of all my dreams*

W.J. Maixner

# Foreword

By C. Sainte-Rose

As we stand at the dawn of the 21st century, one may ponder the rationale of writing a book on spina bifida. Once commonplace in European countries prior to the era of ultrasonography, this disease became increasingly rare in developed countries as a result of improvements in antenatal diagnosis, to the point that we believed it to be disappearing. Knowledge of spina bifida and of its treatment, once so richly diffused only 30 years ago, began to fade. Young neurosurgeons who had never seen such a malformation at its initial presentation were hesitant, and did not understand the protean clinical signs of these patients presenting to the emergency department or outpatient clinics. This situation, however, did not last for long. As a consequence of the political and economic events of the final years of the 20th century, the advent of globalisation, and the significant desire for immigration, we realised that spina bifida had not disappeared at all in the rest of the world. Migration was, and is, bringing it back onto our doorstep, to our everyday clinical and surgical practice. It is important therefore, not to lose the knowledge gained by our masters, to try and assemble it in one place in order to understand the disease from its inception in utero through until adulthood and the reproductive age.

The chronology of the book, in reflection of this aim, is well organised, and ranges from history and embryology, to prenatal diagnosis and treatment, perinatal care, initial treatment and management, and middle- and long-term complications, and finally provides insights for the future. The authors, by not merely focusing on the neurosurgical issues but also on the urologic and orthopaedic consequences of this malformation, have allowed for a more global approach to these patients.

Through a greater understanding of the disease, through the improved quality of initial care, and through the dedication of the surgeons and physicians who manage these children from infancy into adulthood, children inflicted with spina bifida, whilst heavily burdened, may nevertheless lead happy and full lives.

*Paris, May 2008*

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# Foreword

By C. di Rocco

Spina bifida refers to a cohort of pathological conditions that vary in severity from the dramatic and life-threatening myelomeningoceles to relatively mild defects such as dermal sinuses. This book aims to cover the whole spectrum although, as expected, most of its content is devoted to the management of myelomeningocele. With regard to this last type of malformation, the treatment of very few diseases in the history of medicine has been characterized by such an intimate relationship between scientific and technical problems and ethical and social considerations. Myelomeningocele is one of a very few congenital malformations that require the neurosurgeon and parents to face the fundamental dilemma of whether to promote survival, at a very high cost, or to deny treatment in the belief that it is better to prevent the newborn suffering unacceptable and lifelong physical and emotional suffering. It is unusual as it has been associated with such mutually exclusive therapeutic approaches – from treatment refusal to the use of sophisticated surgical and rehabilitative care. The management of the disease has been marked by oscillations between hope and dejection, despair and jubilation. These opposing emotions are also likely to affect the individual surgeon in his/her professional life, with cases of early success turning to late failures and, conversely, cases of satisfactory lives with rich social interaction following initially poor prognoses.

Before the 1960s, in common with other children with hydrocephalus, newborns with myelomeningocele had little hope of survival. Indeed, most of the affected subjects died from the uncontrolled progression of the associated ventricular dilation; death also resulted from sepsis, meningitis, and renal failure. The same secondary complications accounted for the extremely serious disabilities that blighted survivors' lives, in addition to the congenital neurological, orthopedic, and urologic deficits. It is not surprising, therefore, that at the beginning of the 1970s Lorber commented pessimistically on a series of patients followed in the previous decade. He considered that only 7% of the survivors had acceptable disabilities, while the majority of them "had a quality of life inconsistent with self-respect, learning capacity, happiness, and even marriage". A generation of pediatricians as well as the public opinion of the time was deeply influenced by the so-called "Lorber's selection criteria". Macrocrania, severe paraplegia, severe kyphosis or scoliosis, the presence of concomitant significant congenital anomalies, or the history of birth damage were all regarded as criteria for excluding any active treatment for myelodysplasic newborns. The echo of this negative attitude can also be found in more recent experiences, as demonstrated by the "Baby Jane Doe" case at the beginning of the 1980s, and by the "Baby Rianne" case in 1993. In this context, the protocol approved by the Dutch Association of Paediatrics in 2005, known as the Groningen protocol, on deliberate life termination for newborns with severe forms of spina bifida, is even more significant.

On the other hand, although these years were dominated by this negative attitude in most centres, some neurosurgical departments produced results that were much more optimistic. In a small number of neurosurgical centres, series of surgical unselected patients were managed with more advanced techniques and a multidisciplinary approach. The results unequivocally demonstrated that most of the children operated on could reach normal levels of intellectual ability, and that even individuals with severe myelomeningoceles could, after this treatment, lead meaningful lives.

These patients benefited from the surgical advances made in assuring a more effective closure of the back defect. They also had the advantage of improved control of the impaired cerebrospinal fluid (CSF) dynamics by means of more reliable CSF shunting apparatus, and, recently, endoscopy, early recognition and treatment of complications such as Chiari type II malformation or syringomyelia, and prevention and early treatment of spinal cord retethering. They gained further benefit from timely orthopedic correction for club feet, scoliosis, or kyphosis, as well as adequate prevention of damage to urological function. Furthermore, they also received strong support for reaching independence and social integration from the development of ad hoc rehabilitation and educational programs, which were additionally promoted by the establishment of myelomeningocele clinics; more generally, they also benefited from the combined efforts of multidisciplinary teams assisted by a more sympathetic public opinion.

The spirit that has inspired multidisciplinary myelomeningocele teams over the last two decades can be found in this multiauthored volume devoted to the management of spina bifida. The book conveys all the best information currently available in the field, and integrates the most specialized knowledge, from the basic sciences to the various surgical, medical, and psychosocial skills, in a unique well-organized source of information .

As a result of understanding the role of alpha-feto-protein in preventing spina bifida, and the introduction of routine prenatal ultrasound diagnosis, the number of children born with myelomeningocele is continuously decreasing in many western countries. This phenomenon is likely to result in a decreased interest in the management of spina bifida and there will almost certainly be fewer specialized scientific contributions. Consequently, the experience accrued over recent years faces the risk of not being sufficiently updated. Spina bifida, however, still continues to represent an important problem in several countries where preventive measures have not yet been adopted. In these countries, the need for specialized knowledge will persist for the foreseeable future. The present book then may cover an impending gap by helping to preserve the body of relevant scientific knowledge and expertise acquired over the last three decades in a society that may not have so much need of it in future years. By transmitting this skill and knowledge to countries that need it the book will help these countries to avoid retracing the false steps that in the past prevented many children with spina bifida from reaching the best possible outcomes.

*Rome, May 2008*

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# Preface

By M. Necmettin Pamir

This book presents the current understanding, diagnosis and treatment of spina bifida and related pathologies to the neurosurgical literature.

The book has two important qualities: first of all it provides a thorough analysis of the historical experience in the field, and secondly it presents to the reader the most up-to date conclusions, standards, and trends. Although this disease has been known to mankind since antiquity, the treatment is still not straightforward and experts still disagree on various fields. The discussion of modern diagnostic technologies, treatment modalities, and complications, presented in 39 chapters, will guide the reader to tailor an optimal treatment strategy to their patients. The book covers a wide range of topics in the field, starting from preventive measures and progressing to issues like the social adaptation of the patients. The extremely detailed nature of each chapter will be easily appreciated by the reader, whether this is the neurosurgeon, pediatrician, pediatric neurologist, pathologist, neuroradiologist, or any other professional who is involved in the diagnosis or treatment. Therefore, the reader will find answers to almost all his questions on spina bifida and related pathologies in this book. In this regard, it is a significant contribution to the medical literature.

I take great pleasure in congratulating the editors, Professor Özek, Professor Cinalli and Professor Maixner for the quality of the book and for their superb and unique work. It is an honor for me to introduce this excellent book to neurosurgical literature.

*Istanbul, May 2008*

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# Preface

By the Editors

Spina bifida has been an issue of concern for thousands of years. The treatment for this malady begins at birth, or in some cases even before birth, and continues throughout the patients' lives. Over years of investigations and studies in our spina bifida outpatient clinics, we have come to realize the importance of understanding and preparing for the long-term difficulties awaiting our patients. As we are aware that the treatment is multidisciplinary, it is no surprise that the spark of an idea for a spina bifida book was generated during meetings between members of the subspecialties in our spina bifida team. With this inspiration, the decision was shaped further and finalized during the 2006 ESPN meeting in Martinique.

With the aim of promoting the academic success of our book, deciding on the authors for the specific chapters and depending on their expertise was of utmost importance. For this reason, we would like to acknowledge the invaluable contribution of all our authors. We express our heartfelt gratitude to Springer-Verlag Italy, and particularly Dr. Donatella Rizza and the whole editorial team for their skilfulness and tolerance. Lastly, we would like to sincerely thank our patients and their families from whose endurance we get all our clinical experience.

*May 2008*

*M. Memet Özek*
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# Section I
# GENERAL CONSIDERATIONS

# Chapter 1
# A Historical Review of the Surgical Treatment of Spina Bifida

James T. Goodrich

## Spina Bifida in Antiquity

Spina bifida or spinal dysraphisms have been present as long as man has walked the planet. A number of anthropological excavations have uncovered spines with stigmata typically seen in infants born with myelomeningoceles. As these children were born in an era where little or no treatment was available we can only assume that most did not survive. Having said that, there are a large number of surviving anthropological figures sculpted in stone, terracotta and other materials from early civilizations. These sculptures provide evidence of individuals who survived with what would be a normally devastating disease. Over the years the author has collected a number of terracotta figures from the Americas that show clear evidence of surviving children with stigmata of spinal dysraphism. These figures are seated in the typical position of a paraplegic child or adult with the typical lumbar kyphosis. Some of the figures have been incorrectly described as patients with tuberculosis or Pott's disease. A careful examination of these figures clearly shows the physical characteristics of individuals with chronic myelomeningocele. We have included several examples that come from Meso-American cultures where figures of this type are not at all uncommon (Figs. 1.1-1.5).

**Fig. 1.1.** A terracotta figure from Colima, Mexico (circa 200 A.D.), revealing findings classic for a child or adult with a spinal dysraphism. The forward posture with hands resting on the knees and the severe kyphosis of the lumbar spine are classic findings. From the author's personal collection

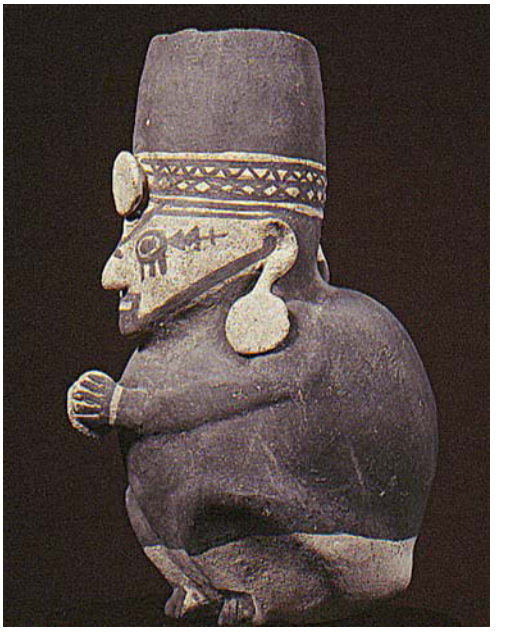


**Fig. 1.2.** A terracotta figure from Chancay, Peru (circa 1000 A.D.), showing an individual with classic findings of spina bifida, including the typical forward posture with hands resting on the knees as a result of paraplegia. The thoraco-lumbar kyphotic spine can be seen in the profile. From the author's personal collection

**Fig. 1.3.** An Olmec child with spina bifida in the typical seating position with severe lumbosacral kyphosis. The child also appears to have hydrocephalus. From the Olmec culture (circa 1500 B.C., Meso-America). From the author's personal collection

**Fig. 1.4.** A terracotta pottery piece from Colima, Mexico (circa 200 A.D.), showing a "shaman" figure with the typical horn on the forehead. The seating position with plegic legs and the lumbosacral kyphosis are indicative of an individual with a spinal dysraphism. From the author's personal collection

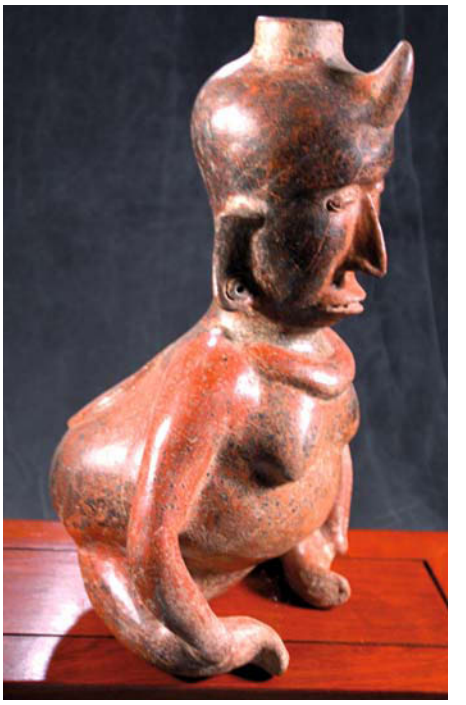


**Fig. 1.5.** An example of a "shaman" represented by the horn on the forehead, a large hydrocephalic head and the typical characteristics of a spina bifida. On the backside, where the myelomeningocele is located, is a large medallion-like character that lies over the severe lumbosacral kyphosis. The patient is sitting in the classic forward pitched position as a result of weak abdominal muscles, with hands resting on the legs – a typical sitting position for a child with a severe myelomeningocele. From the author's personal collection

*"Alius morbus oritur ex defluxione capitis per venas in spinalem medullam. Inde autem in sacrum os impetum facit, quo medulla ipsa fluxionem perdit"*. – Hippocrates [1] (Another disease springs out as an outflow from the head through the veins into the spinal cord. From there moreover it attacks the sacral bone, where the spinal cord itself contains the flow).

Descriptions of what appear to be spinal dysraphisms are found in the early writings of Hippocrates (see quotation above), Galen and others. Review of these early writings indicates that these authors clearly lacked any formal comprehension of the disorder. Surgical treatment for the condition appears to have been virtually nonexistent in the early Greco-Roman era. The earliest definitive description of spina bifida that we have located is that of the Dutch clinician Peter van Forest (1522-1597). In a posthumous work published in 1610, van Forest gave an account of a 2-year-old child with a neck malformation that appears to have been a form of spina bifida. Van Forest surgically ligated the mass at the base but the child went on to die [2].

The first illustrated example of spinal dysraphism appeared in a textbook entitled *Observationes Medicae*, first published in 1641, which went through many editions [3]. The author of this book was Nicholaas Tulp (1593-1674). Tulp (real name, Claes Piereszoon) is best remembered as the main figure in Rembrandt's painting of "The Anatomy Lesson of Dr. Tulp", from 1632, Tulp's second year in practice as an anatomical lecturer. To Tulp we also owe the introduction of the term "spina bifida" [3]. In his textbook, Tulp described six cases, one of which was that of a child with a large lumbar myelomeningocele arising from a narrow pedicle (Fig. 1.6). Tulp described this lesion as "*nervorum propagines tam varie per tumorem dispersas*" (the prolongations of the nerves scattered in different directions through the tumor). For its treatment he described dissecting the myelomeningocele sac and ligating the pedicle; the patient soon died of infection. As a result of this experience, Tulp recommended approaching such lesions with caution as consequences could be dire. Tulp's illustration shows the sac and dissected nerves at autopsy. In reviewing the legend to this plate we find the first printed use of the descriptive term "spina bifida" [3, 4].

To Marco Aurelio Severino (1580-1656), we owe the first textbook on surgical pathology, originally published in 1632 [5]. This work underwent many editions because of its widespread popularity, which is believed to reflect the high quality of the illustrations and the elegant discussions of the case presentations. This remarkable work contains one of the earliest published illustrations of a child with a cervical myelomeningocele (Fig. 1.7). Severino was a widely known and respected teacher in Naples, Italy. He was also an early and important supporter of


**Fig. 1.6.** One of the earliest known printed examples of spina bifida, from Tulp's *Observationes Medicae* [3]. In the legend (*bottom right of image*) of this image is the first use of the term "spina bifida". This diagram was based on a patient in whom, after Tulp attempted to repair the myelomeningocele, sepsis prevailed shortly after surgery. The child also appears to have hydrocephalus, a commonly associated pathological condition

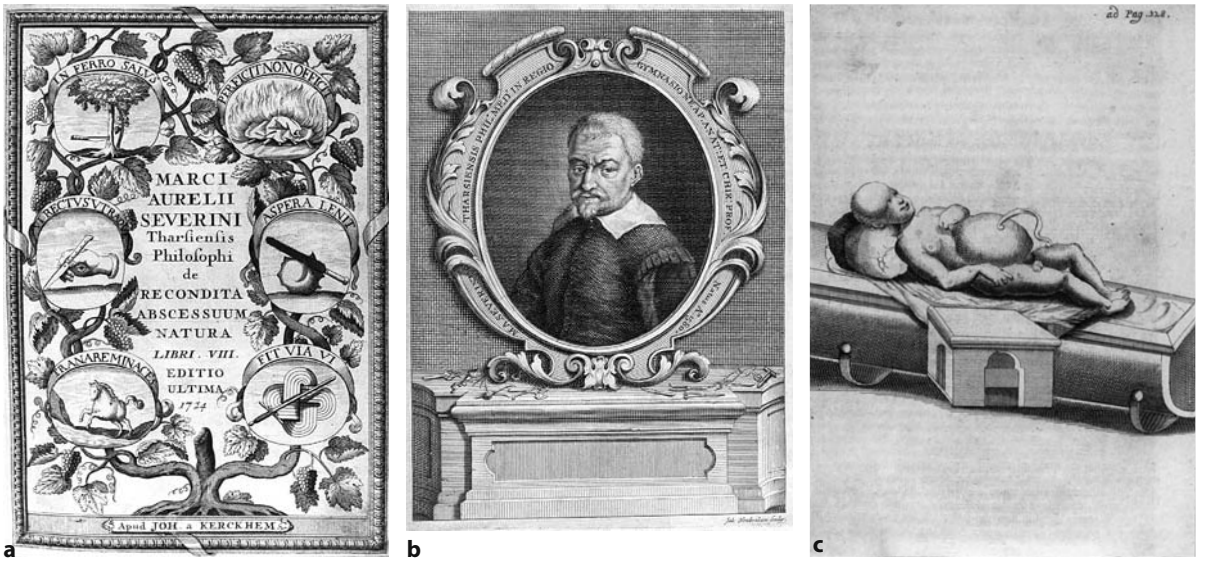


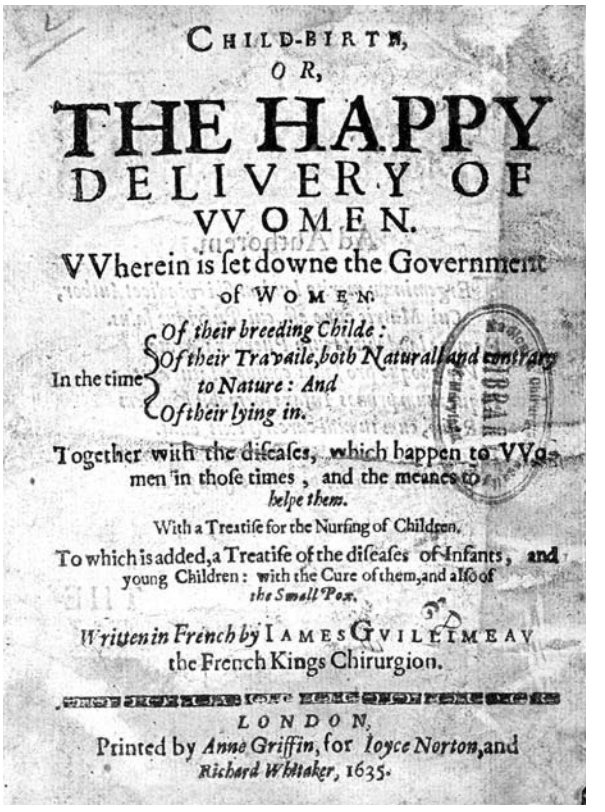

**Fig. 1.7 a-c.** The first textbook on surgical pathology. **a** Titlepage. **b** Frontispiece portrait of Severino. **c** A classic illustration of a child with a cervical myelomeningocele

William Harvey and his ideas on the circulation of the blood. In this book on the "obscure nature of tumors", we find some of the most remarkable and early depictions of pathological lesions and tumors, often called "swellings". In addition, he often added the history of the patient and, if surgery was indicated, the techniques he used. Reflecting a then-prevailing view of spina bifida, however, he rarely considered it as a surgical entity. Even so, his illustration of this disorder is one of the earliest printed examples [5, 6].

Another prominent Dutch surgeon and anatomist, Frederik Ruysch (1638-1731), published the first extensive spina bifida series, ten cases, in 1691 [7]. While Ruysch clearly described the condition, he offered nothing in the way of treatment, as he considered it untreatable. He did, however, associate the pathology of the paralytic limbs with the "want of the spinal medulla". Ruysch was professor of anatomy at Leiden and Amsterdam and is best remembered for his technique of injecting anatomical structures to outline the anatomy. Among his great accomplishments was discovering the concept of fetal nutrition via the umbilical cord. Ruysch gave the following description of spina bifida [8]:

*"A tumor frequently arises in the loins of a foetus, while it is yet an inhabitant of the uterus… If we rightly examine this tumor, it will appear as plain as the Noon Sun to be a dropsy, in part of the spinal medulla and is almost the same disorder, allowing for the difference of situation with that which in the head of the foetus is commonly called an hydrocephalus. Whereas, it is surprising that I should often find the spinal medulla well conditioned below the tumor; whence some children retain the motion of their lower limbs, whereas I have found others with their lower limbs paralytic for want of the spinal medulla. With respect to the cure of this disorder, little or nothing can be done toward it"*.

During this period, a number of midwifery manuals were issued, the most popular of which was a work by Jacques Guillemeau (1550-1613), namely, "*Child-Birth, Or, The Happy Delivery of Women Wherein is set downe the Government of Women in the time of their breeding Childe: of their Travaile, both Natural and contrary to Nature: and of their lying in Together with the diseases"*, published in London in 1635 (Fig. 1.8) [9]. Guillemeau was a prominent Paris surgeon, successor to Ambroise Paré as surgeon to King Charles IX. A review of the text yields only one case discussion of a child with hydrocephalus; no cases of spina bifida were identified, leading one to ask whether this author considered this a "hopeless" disease and hence not treatable.


CHILD-BIRTH,
OR,
THE HAPPY
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Together with the diseases, which happen to VVomen in those times, and the meanes to helpe them.
With a Treatise for the Nursing of Children.
To which is added, a Treatise of the diseases of Infants, and young Children: with the Cure of them, and also of the Small Pox.
Written in French by IAMES GVILLIMEAV the French Kings Chirurgion.
LONDON,
Printed by Anne Griffin, for Ioyce Norton, and Richard Whitaker, 1635.

**Fig. 1.8.** Midwifery manual by Jacques Guillemeau: "Child-Birth; or, The Happy Delivery of Women Wherein is set downe the Government of Women in the time of their breeding Childe: of their Travaile, both Natural and contrary to Nature: and of their lying in Together with the diseases", London, 1635 [9]

To Giovanni Baptiste Morgagni (1682-1771), a teacher of anatomy at Padua, we owe the first solid clinical description of the association of hydrocephalus and spina bifida. In his book on the *Seats and Causes of Diseases*, he gave a clear and vivid description of a child in the post-mortem state who had been born with spina bifida and hydrocephalus [10, 11]. In a section entitled "*Sermo de Hydrocephalus et de Aqueis Spinae Tumoribus*", he discussed several cases of associated hydrocephalus and spina bifida. However, he did not recognize cerebrospinal fluid (CSF) as a physiological entity; rather, he described this fluid collection in the brain and spine as "*hydrops cerebri et medullaris"*, or an excessive collection of fluid. As an anatomist he described only the clinical findings and offered no advice as to how to treat this disorder (Fig. 1.9).

One of the finest and earliest illustrated examples of spina bifida, with hand-colored drawings, was prepared by Jean Cruveilhier (1791-1874), the son of a military surgeon. These important and elegant rep-


**Fig. 1.9.** The second edition of Morgagni's masterpiece "On the Seats and Causes of Disease" with a nice frontispiece engraving of Morgagni – *De sedibus, et causis morboum per anatomen indagatis libri quinque*. Patavii: Sumptibus Remondiananis, 1765 [11]

resentations were published in a series of fascicles published over 14 years, 1829-1842 [12, 13]. Detailed case presentations and clinical findings were included, along with case summaries. Cruveilhier added his own comments on the disease process and observations about treatment. As a result of his considerable clinical acumen Cruveilheir was appointed the first professor of descriptive anatomy (i.e., pathology) at the University of Paris. In addition he worked at the Charité and Salpetrière. In his remarkable two-volume folio work he described a number of significant early pathological conditions. The only criticism of this work could be lack of organization, as a number of unrelated cases were put together in a fascicle. However, it is the only fault that can be found with this remarkable treatise. Among his contributions were a series of illustrations showing dramatic presentations of spina bifida and hydrocephalus. He devoted four sections to these subjects. Two cases of myelomeningocele are discussed along with the clinical course (see Livraison 6, plate 3). One child was seen at three days of age and followed for two weeks when the child died of meningitis. At autopsy Cruveilhier found massive hydrocephalus with flattened gyri and sulci. In addition, there was purulent material in the ventricles that he felt had spread through the subarachnoid space from the spina bifida to the ventricles. He noted that the infection probably coursed through the foramen of Magendie – interestingly, this foramen had only just been recently described.

A second case description of spina bifida is most interesting as Cruveilhier clearly describes what we now call the Chiari Type II malformation nearly 55 years before Arnold and Chiari provide the definitive anatomical description (to be discussed below). The case involved a child with a myelomeningocele who died of sepsis. At autopsy he described the bony anomalies of spina bifida and associated diastematomyelia. The further description of the posterior fossa and cerebellum are typical of what we now call a Chiari Type II malformation. His description: "… the upper part of the cervical region, considerably enlarged, contained both the medulla oblongata and the corresponding parts of the cerebellum which was elongated and covered the fourth ventricle which itself became longer and wider". Cruveilhier described two other cases in which his findings were similar: "… this type of descent of the elongated medulla and cerebellum into the upper part of the spinal canal". Cruveilhier believed that spina bifida occurred secondary to an abnormality of development, in retrospect, a remarkably early insight.

An important further clinical observation by Cruveilhier was that the child with myelomeningocele who did best was the one with a closed non-leaking sac. Once the sac opened, the course was disastrous, with infection, sepsis, paraplegia, seizures and death. He commented on the celebrated case of Sir Astley Cooper, who claimed to have cured a child of this problem by repeated punctures of the sac – his observation was that this was a singular fortunate event and not the rule (see Livraison 16 plate 4) (Fig. 1.10).

A surgeon, M. Baxter, likely unaware of the earlier findings of Morgagni in 1761 and of Cruveilhier, published a note in 1882 in which he described the association of hydrocephalus with meningocele [14]. Another anomaly not uncommonly associated with spina bifida was first described by Lebedeff in a case of spina bifida with anencephaly [15].

In more recent times, anatomical studies have further elucidated spina bifida. Such studies include the classic studies by Kermauner [16], Keiller [17] and Bohnstedt [18]. In an effort to enlarge the concept of spina bifida, Fuchs introduced the term "myelodysplasia" in 1910 to denote spina bifida, enuresis, and associated deformities of the feet [19]. This clinical syndrome was further refined in the classic paper by de Vries in 1928 [20]. Lichtenstein used the term "spinal dysraphism" to describe a pleomorphic group of disorders of cutaneous, mesodermal or neural origin [21]. Lichtenstein was among the first to discuss the neuroanatomic effects of spina bifida on distant parts of the central nervous system [22].

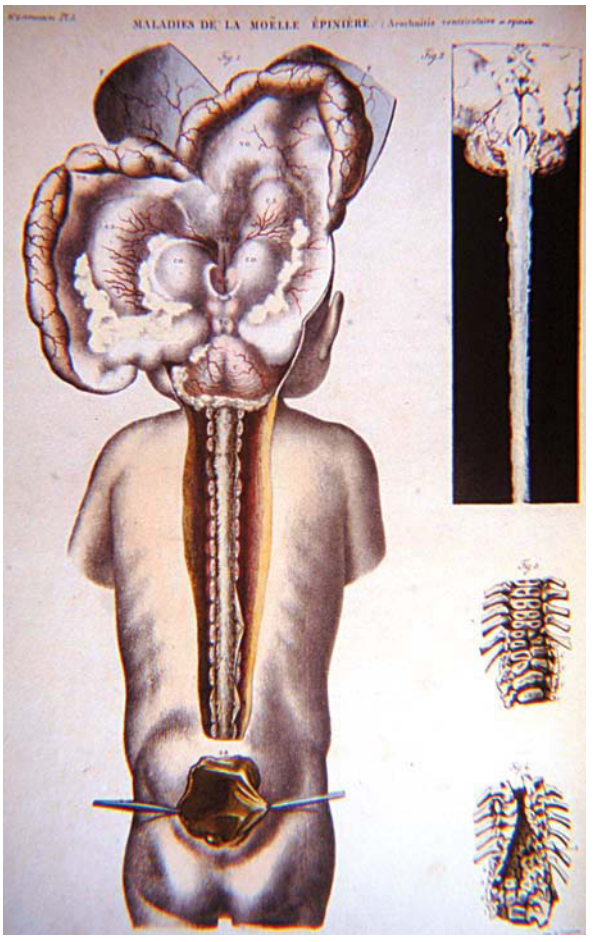


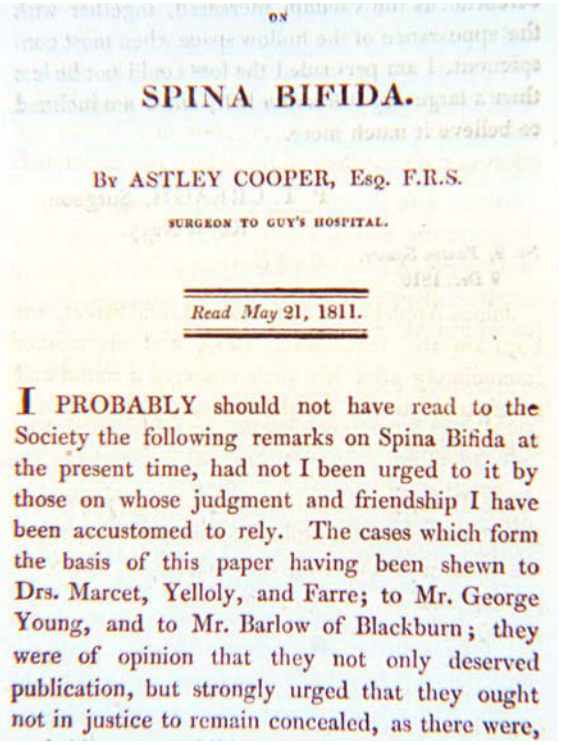

**Fig. 1.10.** An important illustration of a child with a myelomeningocele and hydrocephalus. In the smaller drawings to the right are examples of spina bifida of the spine. Not well appreciated is the tonsillar herniation of the cerebellum at the cervico-medullary junction – the first description of what we now call a Chiari Type II malformation [12]

Review of the surgical treatment of spina bifida over the years indicates that the most common treatment has involved nothing more than ligation or amputation of the sac. The outcome has almost always been fatal, either because of CSF leakage and infection, or the secondary progressive untreated hydrocephalus. Looking back at the literature, we find a typical eighteenth century case of surgical treatment of spina bifida as described by a prominent London surgeon, Benjamin Bell (1749-1806). Bell's treatment was to place a tight snare ligature around the base of the sac and then allow it to slough off. Interestingly, Bell commented that hydrocephalus was not uncommonly associated with myelomeningoceles. The outcome was fatal in all the cases Bell described, leading him to comment: "This is perhaps the most fatal disease to which infancy is liable; for as yet no remedy has been discovered for it… Experience shows, however, that every attempt of this kind should be avoided for hitherto the practice has uniformly proved unsuccessful. The patient has either died suddenly, or in the course of a few hours after the operation" [23].

Sir Astley Paston Cooper (1768-1841), a London Guy's Hospital surgeon well known for his surgical prowess, presented a paper to the Royal College of Surgeons in 1811 on spina bifida [24]. Despite being a skilled surgeon who trained under John Hunter, he summarized a number of prevalent views, all of which culminated in the conclusion that this was an untreatable disease that was best left alone. Cooper had only one successful surgical treatment, a child in whom he performed multiple punctures of the sac. The child's survival represented an amazing feat considering this was in the pre-Listerian era of no antisepsis. In Cooper's view this disorder was only to be treated with measures that are "palliative, by pressure, or curative, by puncture". In his paper he provided a lithograph of the disorder (Fig. 1.11).

Samuel Cooper (1780-1848), a prominent English surgeon and former president of the Royal College of Surgeons, provided an excellent nineteenth century clinical view of spina bifida. In the following statement [25], he clearly summed up the clinical problems related to spina bifida and the contemporary lack of successful surgical treatment: "The generality of children, affected with spina bifida, are deficient in strength, and subject to frequent diarrhoae. Incontinence of urine and the feces, emaciation, weakness, and even complete paralysis, are sometimes the concomitants of this serious complaint. However, some of the patients are, in every respect, except the tumor, perfectly healthy, and well formed". In discussing surgery, Cooper remarked [26]: "Experience has fully proved, that puncturing the tumor with a lancet, and thus discharging the fluid, either at once, or gradually, cannot be done without putting the patient in the greatest danger, the consequences being for the most part fatal in a very short space of time". Surgeons continued to be inventive, offering other techniques for treating spina bifida that included injecting the sac with sclerosing solutions (typically iodine, potassium iodide). Although the reported mortality was less in such cases, nonetheless the frequency of neurological deficits was reported as significantly increased [27].

Following up on earlier injection techniques, Palasciano proposed a "new" method to treat spina bifida cystica and encephalocele in the 1850s [28]. He first emptied the dysraphic sac of CSF, and then



ON

SPINA BIFIDA.

By ASTLEY COOPER, Esq. F.R.S.

SURGEON TO GUY'S HOSPITAL.

*Read May* 21, 1811.

I PROBABLY should not have read to the Society the following remarks on Spina Bifida at the present time, had not I been urged to it by those on whose judgment and friendship I have been accustomed to rely. The cases which form the basis of this paper having been shewn to Drs. Marcet, Yelloly, and Farre; to Mr. George Young, and to Mr. Barlow of Blackburn; they were of opinion that they not only deserved publication, but strongly urged that they ought not in justice to remain concealed, as there were,

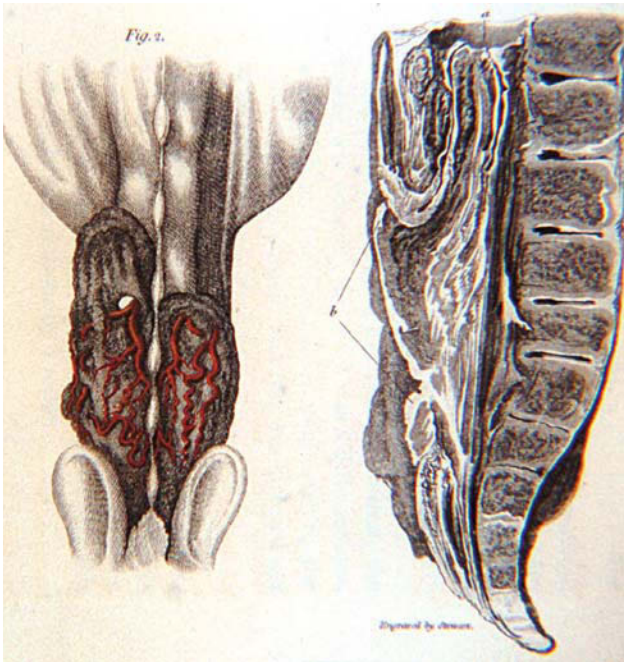



**Fig. 1.11.** Title page and illustration of spina bifida from Astley Cooper's monograph on spina bifida [24]

concentrically compressed the sac to bring together either the cranial or the vertebral margins of the defect. He then injected iodine into the sac to induce sclerosis. A surgical technique for myelomeningocele was further refined by Francesco Rizzoli (1809-1880) in 1869 [29]. He abandoned the technique of using sclerosing iodine as he felt it was too damaging to the nervous elements. As an alternative method he designed and applied a "Rizzoli enterotome" to the dysraphic sac and slowly closed it, allowing the sac to necrose and slough off (Fig. 1.12).

In the nineteenth century a number of atlases were produced that dealt with "human monsters" and a number of these illustrated various cases of myelomeningoceles. One of the most popular atlases was by Friedrich Ahlfeld, entitled *Atlas zu die Missbildungen des Menschen*, published in Leipzig in 1880-1882 [30]. The work was on a number of human malformations with some elegant lithographic plates. Some examples of spina bifida from this volume are included (Figs. 1.13-1.15). Ahlfeld felt this disorder was due to an excess collection of fluid, not recognizing the physiology of CSF: "*Man muss für die Mehrzahl der Fälle die primäre Ursache im spinalen Hydrops suchen*". There is no discussion of surgical management in this work. It was not until the work of Lebedeff (1881-1882) that the concept was put forward that spina bifida resulted from a failure of neural tube closure in early fetal development [15]. Lebedeff commented on this disorder as "*Entstehung der Hemicephalie und Spina Bifida zurück auf Anomalie Krümmungen des Medullarrohrs in der frühesten fötalen Period*" [31].

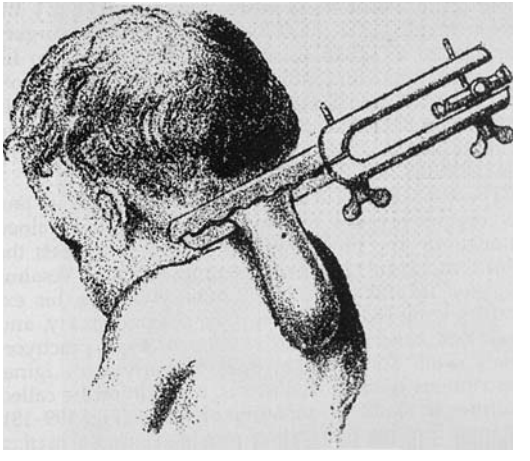


**Fig. 1.12.** An example of the "Rizzoli enterotome" for removing a myelomeningocele. The instrument was applied and then "slowly" closed, causing the sac to necrose and eventually fall off

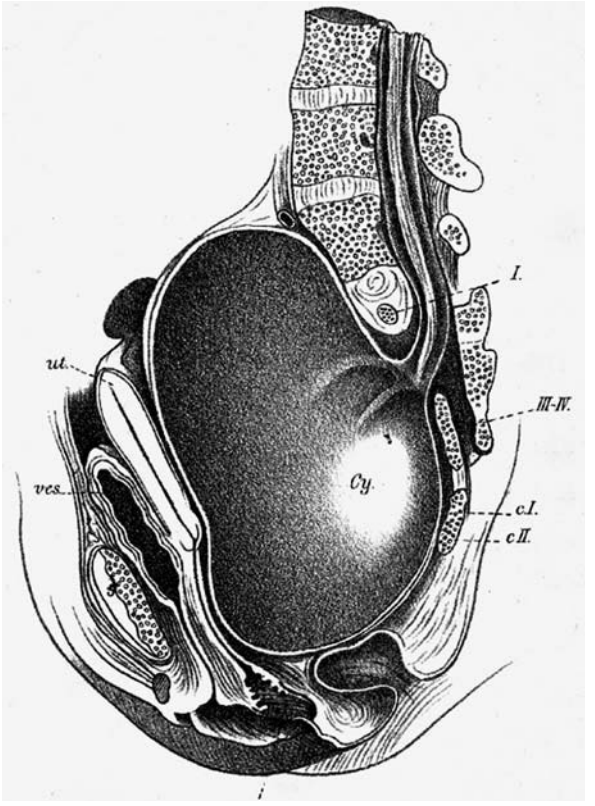



**Fig. 1.13.** From Ahlfeld's *Atlas zu die Missbildungen...* (1880-1882) showing a nice example of a rare anterior myelomeningocele

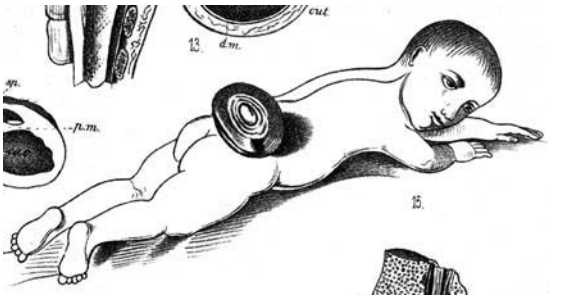



**Fig. 1.14.** From Ahlfeld's *Atlas zu die Missbildungen...* (1880-1882) illustrating a newborn child with a large untreated myelomeningocele

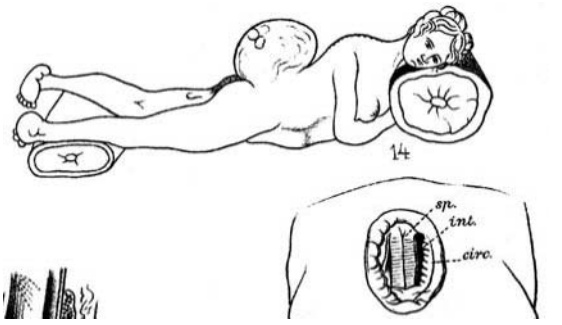



**Fig. 1.15.** From Ahlfeld's *Atlas zu die Missbildungen...* (1880-1882) showing a nice example of an adult female with a large untreated myelomeningocele with clinical lower extremities weakness and deformation

One of the most important nineteenth monographs on spina bifida was published by Friedrich von Recklinghausen (1833-1910) [32]. Von Recklinghausen was a pupil of Rudolf Virchow (1821-1902) at the Pathological Institute in Berlin, Germany. Often forgotten is the fact that it was Virchow who coined the term "spina bifida occulta", now part of our standard nomenclature [33]. Von Recklinghausen eventually settled in Strasbourg, where he remained for the rest of his career, producing his monograph on spina bifida in 1886 [32]. He is probably better remembered for his work on neurofibromatosis, now called "von Recklinghausen disease". In his monograph he described a remarkable case of an adult with spina bifida occulta and hypertrichosis. The patient had a club foot that lacked sensation because of the spina bifida. An ulcer of the foot developed and became septic and the patient died from septicemia. At autopsy, von Recklinghausen found an occult spina bifida of L5 into the sacrum. The conus medullaris was tethered at S2 with a "fatty tumor", likely a lipomyelomeningocele. He made some interesting pathological postulates in that he felt the lipoma was due to abnormal separation of the mesoderm during early embryological development. Von Recklinghausen made a number of other important points in that he appreciated that the fluid in spina bifida came from the subarachnoid space, i.e., it was CSF. He also observed that some patients with spina bifida survived into adulthood as functioning individuals. He noted that hydrocephalus was not always associated with spina bifida. Included in this work are some remarkable illustrations, including two folding plates that clearly outline both the internal and the external pathology of spina bifida. In the monograph von Recklinghausen presented a clear, detailed analysis of the formation of the neuroaxis in myelomeningoceles and spina bifida (Figs. 1.16, 1.17).

The concept of treating a myelomeningocele with a "sclerosis" injection was an old one, but was further embedded in the surgical literature by Morton in 1877 [34]. Morton felt that the surgical outcomes of ligation, amputation, and related procedures in relation to myelomeningoceles led to unacceptable outcomes. He therefore devised a solution that consisted of iodine in glycerine for injection into the "tu-

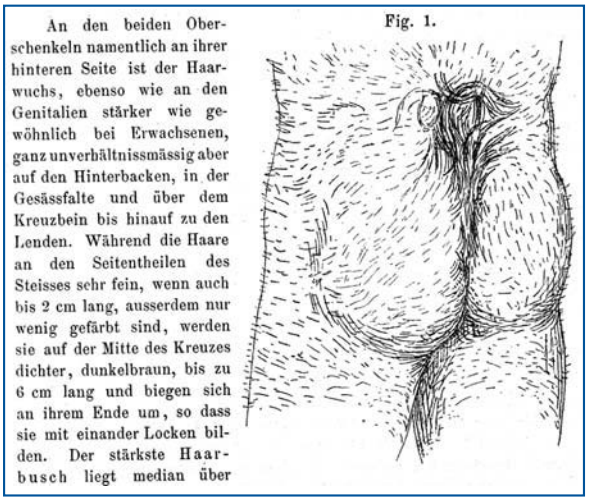

An den beiden Oberschenkeln namentlich an ihrer hinteren Seite ist der Haarwuchs, ebenso wie an den Genitalien stärker wie gewöhnlich bei Erwachsenen, ganz unverhältnissmässig aber auf den Hinterbacken, in der Gesässfalte und über dem Kreuzbein bis hinauf zu den Lenden. Während die Haare an den Seitentheilen des Steisses sehr fein, wenn auch bis 2 cm lang, ausserdem nur wenig gefärbt sind, werden sie auf der Mitte des Kreuzes dichter, dunkelbraun, bis zu 6 cm lang und biegen sich an ihrem Ende um, so dass sie mit einander Locken bilden. Der stärkste Haarbusch liegt median über

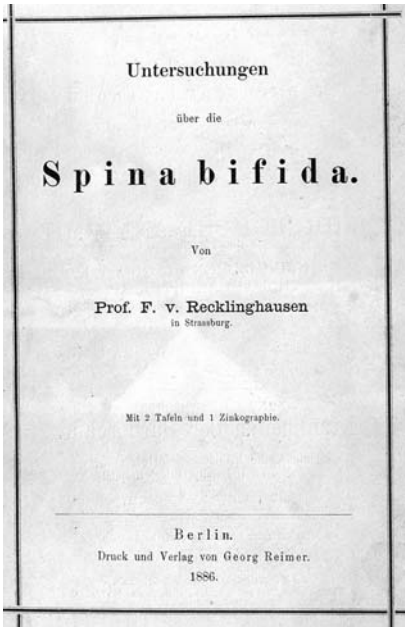
Fig. 1.

**Fig. 1.16.** An illustration of the adult case that von Recklinghausen presented with a lipomyelomeningocele, spina bifida, and hypertrichosis [32]

mour". This technique became quite popular and was used throughout the United Kingdom and Europe.

A late nineteenth century advocate for surgical closure of myelomeningoceles was a Boston surgeon, Henry O. Marcy. In a paper published in *Annals of Surgery* in March of 1895, he came out strongly in favor of surgical repair, in order, so to speak, to create in the closure what nature had failed to do [35]. He felt that failure to do so led to "sepsis *in loco*" and the death of the patient. His argument, and a legitimate one, was that "A priori reasoning would lead to the conclusion that art could supplement nature in her defective development". The case report he presented was that of an 18-year-old female born with a "soft swelling under the skin in the lumbar region". Because the swelling's steady growth had recently become more rapid, resulting in thinning of the sac with the danger of rupture, he decided on surgical intervention. Clinically she had only club feet and was otherwise normal. She "has consulted many surgeons, who have invariably advised against surgical interference". At surgery, the sac contained "one gallon of perfectly clear colorless fluid, which was drawn off by means of a trocar". It is interesting to note that he did the operation on October 16, 1895 and wrote his report and published it on November 9, 1895! He noted that the patient was still in bed with her only complaint that of burning in the feet. Marcy then comprehensively reviewed the nineteenth century surgical literature. He became a clear advocate of surgical closure, as pressure, sclerosis, taps and the like all led to mostly bad outcomes.

In England, a surgeon by the name of J. Cooper Forster presented a case of a child with lumbo-sacral myelomeningocele [36]. Included in his book was an


Untersuchungen

über die

Spina bifida.

Von

Prof. F. v. Recklinghausen

in Strassburg.

Mit 2 Tafeln und 1 Zinkographie.

Berlin.

Druck und Verlag von Georg Reimer.

1886.

a

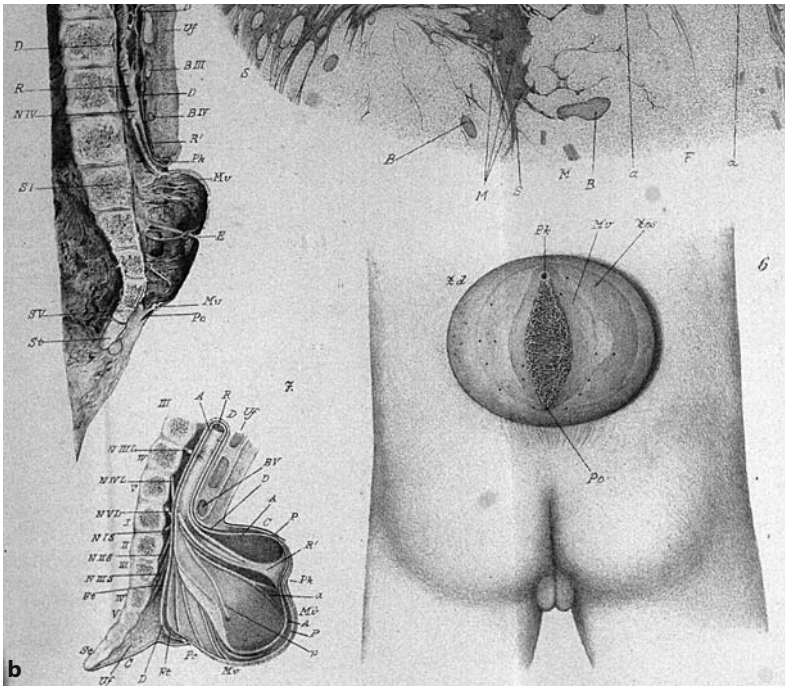

b

**Fig. 1.17 a, b.** **a** Title leaf from von Recklinghausen monograph [32] on spina bifida (from the author's personal collection). **b** Illustrated examples of spina bifida from von Recklinghausen

elegant hand-colored engraving of the lesion. The treatment he advocated was compression of the sac with gentle tapping off of the spinal fluid (Fig. 1.18).

Review of a prominent textbook of surgery from 1887, by John Wyeth of New York, reveals that the author was intrigued to see how little was offered in surgical treatment of myelomeningocele [37]. In most cases treatment was only palliative, with compression of the sac. In the large sac, tapping off of fluid was feasible, but required care and avoidance of the midline because of the nearby neural elements. Wyeth cautioned that the needle should always enter the sac from the side. The smallest needle should be used and only small quantities of fluid taken at any one time. In some cases, Wyeth suggested the injection of Morton's solution into the sac – a commonly recommended practice in the latter half of the nineteenth century. In discussing prognosis of patients with spina bifida, Wyeth commented, "The prognosis is, as a rule, very unfavorable".

Following a similar theme in treatment were the surgical thoughts of Roswell Park, whose volume on surgery was a popular text at the end of the nineteenth century [38]. In the chapter on spina bifida is a clear description of its various types, including illustrations of the anatomy and case presentations. In discussing treatment, one could be either "conservative or operative". Conservative treatment involved only tapping the sac, use of compression or injection of the ubiquitous Morton's solution. An additional thought in treatment was the addition of collodion and iodoform to the puncture site to help reduce further leakage and potential infection. Roswell Park's surgical technique involved freeing the neural elements and dropping them back into the canal. A wide-undermining of the skin flaps gave a primary closure over the defect. He also discussed the use of osteoplastic flaps to close the defective lamina. He felt this is a most difficult maneuver and did not recommend it. Park clearly stated that surgical intervention for spina bifida had only recently become possible with the introduction in the 1870s of aseptic techniques, since before this outcomes were almost always fatal because of septicemia (Fig. 1.19).

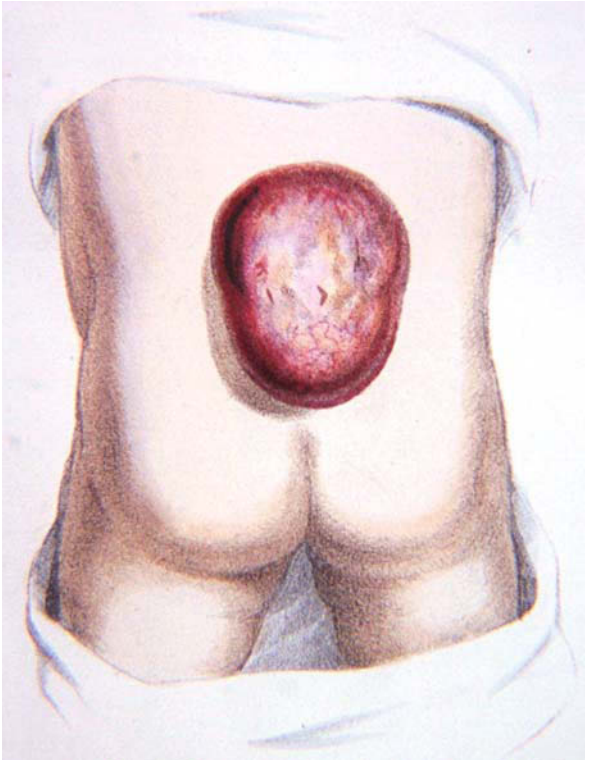


**Fig. 1.18.** From Forster monograph on surgical diseases of children [27]. A classic example of a lumbo-sacral myelomeningocele

In his monograph on *Surgery of Childhood*, Sidney Wilcox, a New York homeopathic surgeon, summarized the surgical treatment of spina bifida at the turn of the century [39]. Wilcox felt this was a disorder that should be approached with some trepidation, as most authors had reported a dismal outcome when spina bifida was treated surgically. Techniques advocated at that time included injection into the sac of Morton's solution [34], which he described as a mixture of iodine, Kali iodide and glycerine. Other techniques included draining the sac and then "darning across the opening with silver wire". Wilcox described a successful case where "the tumour was removed, the laminae were united and the cord sutured end to end". But with some conservative insight he recommended that in those cases where surgery could not be done, to make a small wire cage and fit it over the myelomeningocele, which should be "… filled with cotton so as to make pressure and at the same time ward off blows, [and would thus] be of service" (Fig. 1.20).

The treatment and coverage of open myelomeningocele defects by rotating various skin and muscle flaps were first introduced in 1892, when a German surgeon, C. Bayer, reported on surgical repair of an open myelomeningocele using a series of rotating flap techniques [40]. This surgical technique introduced a new and important concept of placing the neural elements within the spinal canal and then covering the spinal elements with layers of surrounding tissues – a remarkable advance! (Fig. 1.21c).

Antoine Chipault (1866-1920) of Paris, France, prepared a number of important monographs on surgery. Of particular interest is his 1894-95 two-vol-

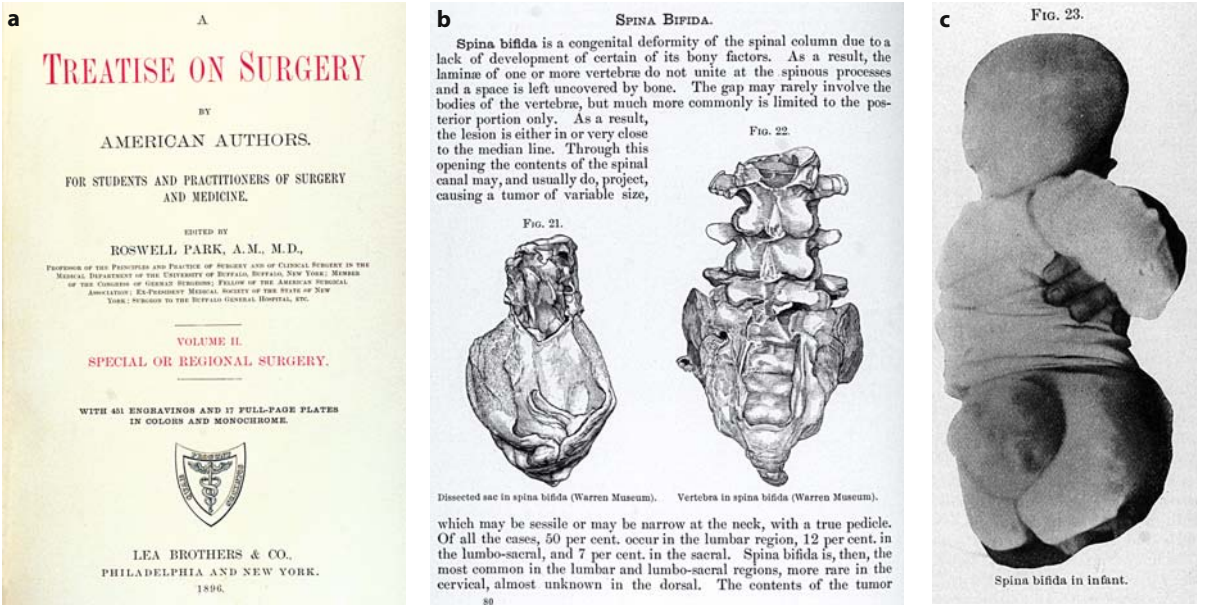



**Fig. 1.19 a-c.** Illustrations from Park's work on surgery. **a** Title page. **b** The anatomy of spina bifida. **c** A clinical example of a child with spina bifida [29]

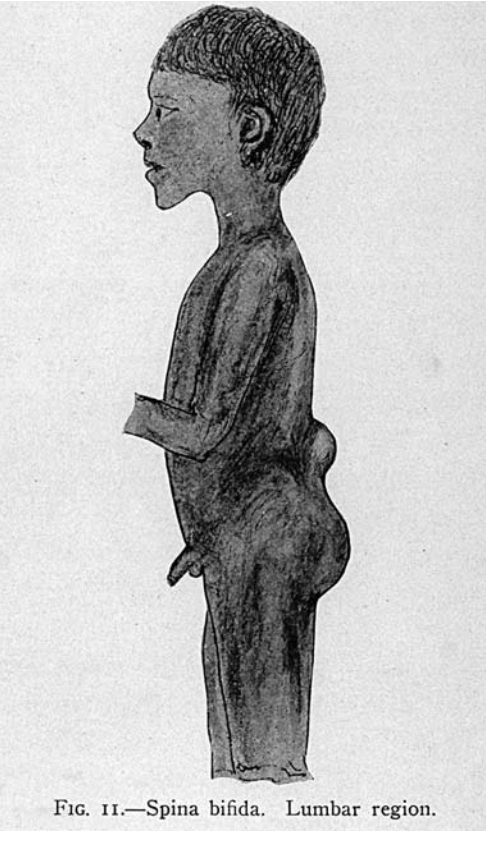



**Fig. 1.20.** A child with an untreated myelomeningocele from Wilcox's *Surgery of Childhood*, 1909. In this case a "wire cage" would be placed over the lesion, filled with cotton to protect it from outside blows [30]

ume work entitled *Chirurgie Opératoire du Système Nerveux*… [41]. In this work he presented a number of examples of spina bifida along with a surgical treatment plan (Fig. 1.20). As was typical throughout the work, the anatomy and the surgical concepts were both impeccable and surgically insightful. Chipault became one of the first European surgeons to argue for a multi-layer closure of the myelomeningocele, adopting the techniques of Bayer [4]. Using aseptic techniques, just recently introduced, Chipault had reasonably good outcomes. Illustrations from his textbook showing the anatomy and techniques for a repair of a myelomeningocele are shown in Figure 1.21.

To Hans Chiari (1851-1916) we owe the development of the anatomical understanding of the "Chiari malformation", often seen in myelomeningoceles, among other disorders [42-46]. A skilled anatomist and pathologist, Chiari is credited with performing over 30,000 autopsies using the systematic autopsy techniques of Karl von Rokitansky (1804-1878) under whom he trained. Often forgotten is the fact that Chiari published the first case of traumatic pneumoencephaly, a publication that later influenced Walter Dandy's development of the pneumoencephalogram [47]. However, it is Chiari's studies on congenital brainstem anomalies associated with hydrocephalus that remain classic [42, 43]. On the basis of his work at the Kaiser Franz Joseph Children's Hospital in Prague he described three different malformations found in 63 patients with hydrocephalus.

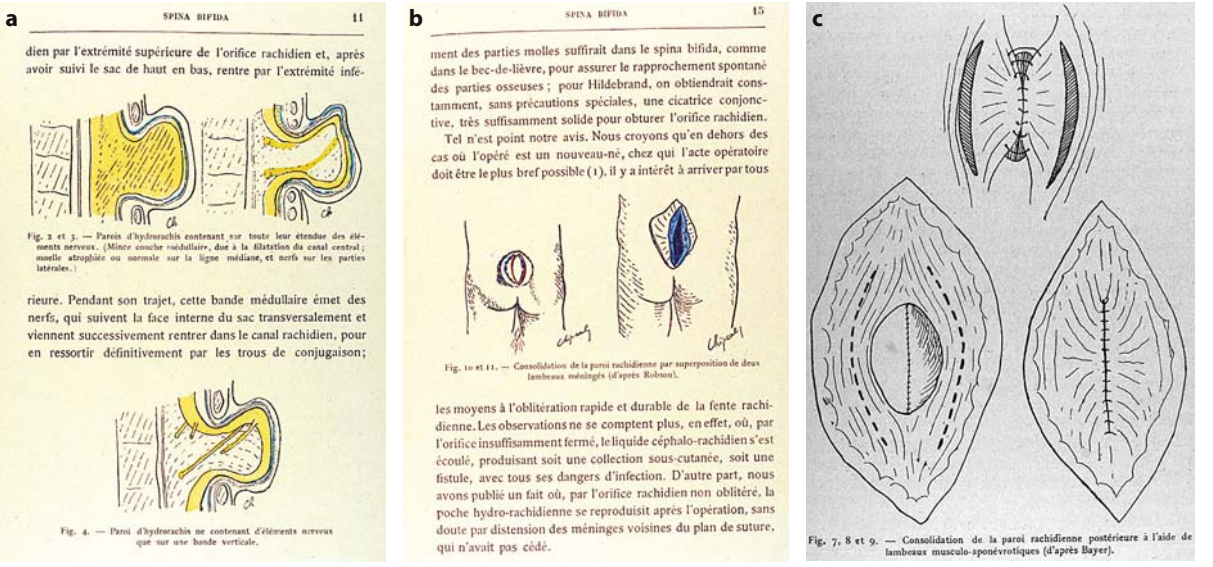



**Fig. 1.21 a-c.** Images from Chipault's textbook [41]. **a**, **b** The typical anatomy of the spinal myelomeningocele. **c** The repair of a myelomeningocele in a multi-layer closure is shown. Chipault adopted this technique from Bayer [40]

Currently we divide these hindbrain anomalies into four types, but the one germane to this paper is the type 2 malformation. Chiari described 14 cases of the type I malformation in patients aged 3 months to 68 years. Interestingly, all of the patients were adults and had hydrocephalus and only one had a myelomeningocele. He called this a deformity of the brain stem that resulted in hydrocephalus: "*Über Veränderungen des Kleinhirns infolge von Hydrocephalie des Grosshirns*". Chiari's type II malformation was described in seven cases in patients ranging from the ages of birth to 6 months. The combination of hydrocephalus and spina bifida was seen in all seven cases. The anatomical findings were a prolongation of the cerebellar vermis and the tonsils (as seen in type I). Associated with this was an inferior prolongation of the fourth ventricle into the cervical spinal canal. Often seen was a kinking of the inferiorly displaced medulla oblongata. In type III there was inferior displacement of the brainstem with herniation of the cerebellum and myelomeningocele. The fourth ventricle was dilated. Type IV was described as cerebellar hypoplasia. Interestingly, Chiari had only one case of type III and described no cases of type IV. Type IV per se was not described until a later monograph in 1896 [43].

For the record, priority in the description of the Chiari type II malformation does not go to Chiari. Rather, this description was first published by John Cleland in 1883, some eight years before Chiari [48]. Chiari gave credit to Cleland's earlier description in his first monograph [42]. Cleland was a Scottish surgeon and anatomist teaching at the University of Glasgow and based his findings on autopsies of nine infants. One of the cases was a child with spina bifida and hydrocephalus and findings that we would now call a Chiari type II.

Julius Arnold (1835-1915), whose name is often associated with this anomaly, published his findings on brain stem anomalies in an infant with multiple congenital anomalies including a large thoraco-lumbar spina bifida in 1894 [49]. Interestingly, the child described did not have hydocephalus. He attributed this anomaly to a primary disturbance in the organization, or disorganization, of the germ cells – a concept he called "monogerminal teratomatous malformation". Two pupils of Arnold, E. Schwalbe and M. Gredig, published a paper in 1907 discussing the association of the hindbrain anomalies with spina bifida and coined the term "*Arnold'sche und Chiari'sche Missbildung*", which led to the "Arnold-Chiari malformation" [50]. Most modern writers have given priority for these descriptions to Chiari and hence the current use of "Chiari" when describing these various malformations [44-46].

By the early part of the twentieth century surgeons were developing better surgical concepts of the repair of myelomeningoceles, i.e., multilayer closures using dura, fascia, muscles and skin. A good example of these newer concepts appears in a classic textbook on the spine by Charles H. Frazier (1870-1936) [51]. Frazier was professor of surgery in Philadelphia and one