Posterior Fossa Tumors in Children

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To my wife for 32 years, Eren, for her support and patience throughout my life.

M. Memet Özek

To my mother.
To Fabrizia, Francesco, and Maria Allegra because the time spent with them is the only real happiness.
and
To Roberta Migliorati who devoted her whole life to the care of children affected by Brain Neoplasms.

Giuseppe Cinalli

To my mentors and students and the children for whom we care.

Wirginia Maixner

To all the young colleagues whom I have helped to progress in pediatric neurosurgery.

Christian Sainte-Rose

Preface

Tumors of the posterior fossa are one of the most challenging pathologies a neurosurgeon is called upon to deal with. The anatomical complexity of the region and the amazing variety of possible histologies of neoplastic lesions in this area in children make the therapeutic challenges even more difficult for the pediatric neurosurgeon. The frequent association with hydrocephalus and the options for its management before, during, or after the surgical procedure on the posterior fossa have given rise to significant controversies during recent years, and a consensus is still far from being obtained. During the last few years, we have witnessed impressive progress in the genetic and genomic profiling of some tumor lesions, allowing the identification of specific and very different prognostic subgroups previously labeled with the same name and often treated with the same protocols. With this new approach, we are entering an era in which we shall be able to tailor treatment protocols very precisely in order to avoid unnecessary procedures or therapeutic regimens, thus limiting the possible collateral effects that have always burdened the long-term prognosis and quality of life of survivors.

We have tried to group into different sections the main pathologies encountered in this age range. For each pathology, recognized experts thoroughly analyze all aspects of genetics, radiology, surgery, pathology, oncology, and radiotherapy. Although all of the editors are surgeons, only Section II is dedicated solely to surgical approaches and techniques, and a strong effort was made when profiling the book plan to offer a real multidisciplinary view of these pathologies. We hope that the final results will reflect this effort. Treatment of posterior fossa tumors in children is never a single person's work. Classification is complex, deeper expertise is demanded of actors in many different fields, and very strong and reliable teamwork is not simply an option but a real obligation.

An entire section has been dedicated to rare pathologies where early recognition may modify the therapeutic approach from the earliest stages, and the final section is devoted to an analysis of different standards of immediate postoperative care and the long-term general implications of follow-up and treatment.

The final result explains why gestation was long and complex, and we are very grateful to all contributors for their patience and to the Springer editorial staff, who believed in this project.

The final acknowledgment always goes to our patients and to their families, who are called upon to face something bigger than themselves and from whose terrible endurance and tribulations the cold scientific aspects of these pages are derived.

Istanbul, Turkey — M. Memet Özek
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General Principles of Treatment in Pediatric Posterior Fossa Tumors

History of Posterior Fossa Tumor Surgery

1

James Tait Goodrich

1.1 Introduction

Development of surgical techniques for the treatment of posterior fossa disorders is, in the terms of historical events, very recent. In the sense of "modern" surgical techniques, we are only looking at a period that developed in the late quarter of the nineteenth century. In looking back at the historical literature, surgeons since the time of antiquity avoided any kind of surgical intervention within the posterior fossa. Early surgeons quickly realized that this region of the brain is extremely sensitive to any type of manipulation. Loss of respiration, sudden death, and distortion of the brain stem, all could lead to a rapid demise of the patient. As we shall see, surgery of the posterior fossa really only came in being with the origins of the twentieth century. A review of surgical textbooks in the latter half of the nineteenth century reveals only a minimal discussion of surgery in the posterior fossa. In the last 25 years, there has been a virtual explosion of techniques and equipment related to what is now called frameless surgical technology. To provide a historical perspective on this subject, the author will review the development of posterior fossa surgery with only a brief look at antiquity and then quickly move to the Renaissance and the pioneering work of the anatomists of the sixteenth century. This chapter will review how the anatomy of the posterior fossa was first understood. The evolution of surgical technique and the individuals who provided us with these new ideas will be discussed.

In the twenty-first century, neurosurgeons now enter an operating room with an environment much more advanced and technically more complex than what was available to their colleagues just 30 years ago. In the 1970s, the surgical microscope revolutionized operative approaches with much improved visualization assisted by better illumination, and as a result, surgeons could now operate more safely on areas of the brain previously considered unapproachable. Looking to the future, likely within the next decade, with the advance in computer-assisted devices, the "hands-on surgeon" will become a relic of the past. The next generation of neurosurgeons will be "data suppliers," i.e., a technician who makes entries into a database and then sits back and watches the robotics, e.g., the "da Vinci" perform the surgical operation.

Having said that, we have made enormous progress in surgical technique and the operative management of patients with complex posterior fossa disorders. To provide a comparison, I would like to present a scenario of a 1930s era operating room, what was then considered a "modern" operating theater. Paul Bucy, a pioneer in American neurosurgery, described the following

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scene at the Queens Square Hospital in London, where he was working as a visiting surgeon:

> …[A]t the National Hospital were Sir Percy Sargent and Mr. Donald Armour. They were both poor surgeons, unbelievably crude in their surgical procedures. On one occasion (Gordon) Holmes told me to go with a patient to the operating theatre and tell Sargent that because the lesion was probably an arteriovenous malformation, he should use great care in exposing it. I did tell Sargent, but he paid no attention and proceeded to open the dura mater with a pair of sharp pointed scissors. In doing so he ripped the malformation wide open, resulting in a severe hemorrhage and the patient's death. On another occasion Armour performed an occipital craniectomy with hammer and chisel. This patient also did not survive the operation. There was a story current at the National Hospital that Denny-Brown, then a house officer, when assisting Armour in an operation would often remark that the blood had reached the drain in the floor on the far side of the room and that perhaps it would be wise to terminate the operation. [1]

This historical vignette clearly shows how far we have come in a fairly short period of time. A more complete anatomical foundation with a better understanding of the disease processes combined with computer technology has clearly led to better surgical outcomes and results in treating disorders of the posterior fossa. Understanding how we have managed to get to this point over time makes for an interesting historical review of our field.

This review will trace the origins of posterior fossa surgery from its antecedents in the Renaissance to the 1940s. Unfortunately, only a sampling of themes and personalities can be provided due to page constraints. Other authors in this monograph will deal with the developments in posterior fossa surgery from the 1940s forward.

1.2 Antiquity

Surgical operations on the posterior fossa have been dated back to antiquity [2]. From skulls excavated from around the world are examples of trephinations of the posterior fossa, most of which were done for trauma or other unknown reasons. In my own collection is a skull obtained from Peru in the 1950s that originates from a graveyard that dates back to 600 A.D. In the mid-portion of the suboccipital bone is a large trephination with healed margins indicating the individual survived the surgical procedure. The reason for the trephination is not clear and it does not appear that it was done for trauma. In reviewing a number of trephined skulls over the years, the vast majority was performed over the convexity, and trephinations of the posterior fossa are actually quite rare Fig. 1.1.

The only accounts of posterior fossa surgery from the Greco-Roman era appear in the writings of Galen of Pergamon (130–200 A.D.), from an area of what is now Turkey [3–6]. In Galen's writings on anatomical procedures, he describes a series of animal dissections in which he exposed the cerebellum and the fourth ventricle, investigations which were done in the second century A.D. Using primarily the rat, he made a linear incision from the inion down to the foramen magnum. Galen studies were done in living animals with bleeding being controlled by finger pressure and scalp retractors. The craniectomy was done with a series of chisels, especially designed for this operation. After the skull bone was removed, Galen described the pulsating brain, especially seeing it rise up out of the craniectomy when the animal was agitated. Galen's technique of opening the dura was no different than what we do today. Galen used a small hook to elevate the dura away from the cerebellum and then incised it with a sharp knife carefully avoiding any of the venous sinuses, overlying cortical vessels, and the cerebellum. Galen pointed out in his discussion of the surgery that problems like cessation of breathing could occur along with motor or sensory loss. The voice could become hoarse and even death could occur. From experimentation, Galen noted that compression of the fourth ventricle could lead to severe impairments and even death. It is well known that Galen was a surgeon to the gladiators, so it is possible that he was involved in the treatment of traumatic injuries to the posterior fossa; whether he ever operated on these types of injuries is open to conjecture. Interestingly, Galen describes in his anatomical dissections splitting the vermis to expose the fourth ventricle in living animals. Galen carefully adds his comment that severe neurological impairments can happen with this technique; nevertheless, it was a useful way to

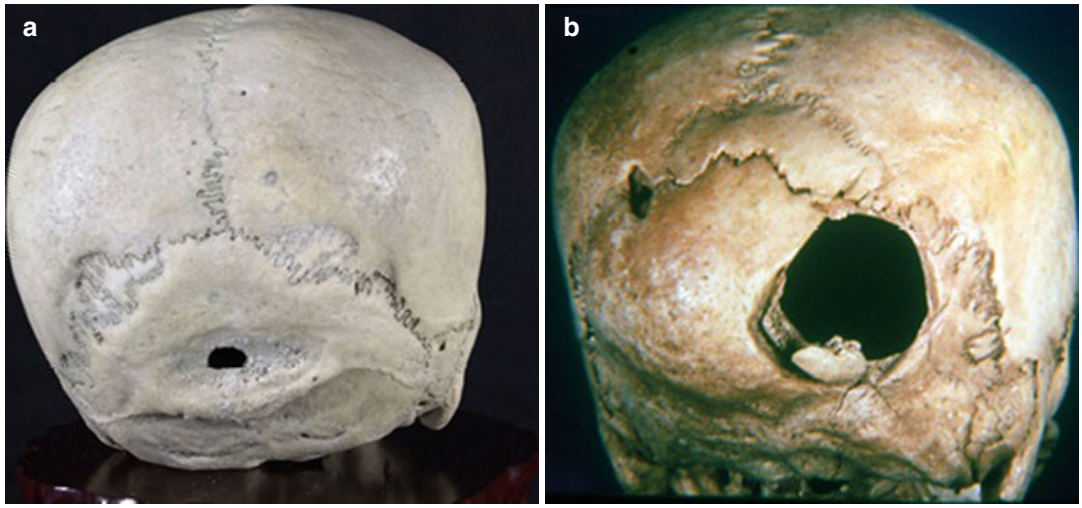

Fig. 1.1 (**a**) A human skull from Peru dating to about 600 A.D. In the occipital bone is a trephination done for unknown reasons. The well-healed bony margins indicate the "patient" survived the operation. From the author's collection. (**b**) A skull with a large right occipital trephination done for unknown reasons. Along the lower edge of the trephination are signs that show healing, so it is likely the patient survived this procedure. Courtesy of the Museum of Man collection, San Diego, California

expose the floor of the fourth ventricle. Galen was the first to describe the calamus scriptorius, which is seen on the floor of the fourth ventricle using this surgical exposure. Galen, throughout his writings, noted that knowledge of the surgical anatomy was absolutely key for the surgeon; without this knowledge, the surgeon would be prone to serious errors and bad outcomes. Unfortunately, much of Galen's animal anatomy was incorrectly transliterated into human anatomy and then carried forward by various translations (e.g., Latin, Greek, and Arabic) to the time of the Renaissance. As we shall see, it was the Renaissance artist that led the drive to first understand and describe human anatomy from hands-on dissections of humans Fig. 1.2.

1.3 The Sixteenth Century: The Origins of Modern Anatomy and Surgical Investigation

At the end of the fifteenth century, the intellectual currents in Europe were undergoing profound changes. With the introduction of the printing press and moveable type, books could be more easily and cheaply produced. As the intellectual shackles of the Middle Ages were

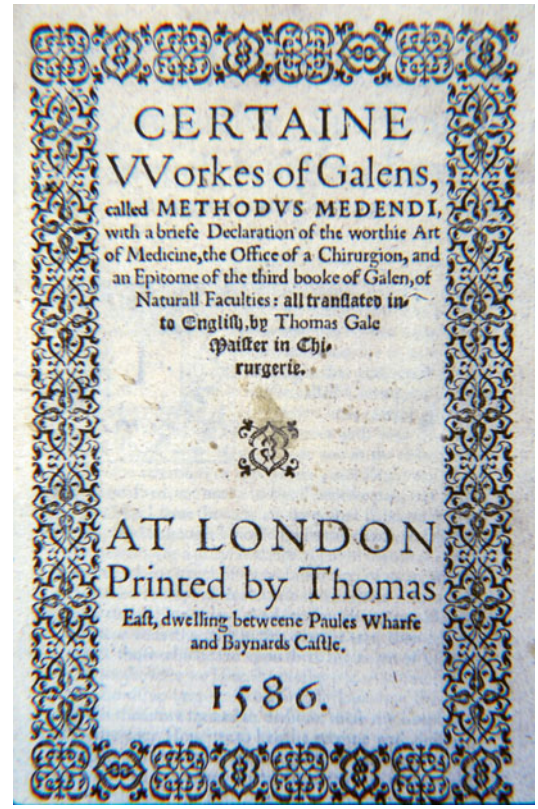
CERTAINE
VVorkes of Galens,
called METHODVS MEDENDI,
with a briefe Declaration of the worthie Art
of Medicine, the Office of a Chirurgion, and
an Epitome of the third booke of Galen, of
Naturall Faculties: all translated in-
to English, by Thomas Gale
Maister in Chi-
rurgerie.
AT LONDON
Printed by Thomas
East, dwelling betweene Paules Wharfe
and Baynards Castle.
1586.

Fig. 1.2 An early English translation of Galen's writing including his "office of a chirurgion" in which he details his surgical techniques along with general works on medicine [5]

being removed, physicians were beginning to rely more on what their eyes taught them at the bedside. The previously held concepts of the early anatomists like Galen of Pergamon and others would be challenged in their accuracy. One of the most important intellectual currents in surgery and medicine at this time were the schools of anatomical studies like Michelangelo, Titian, and Leonardo da Vinci among others [7, 8]. In an attempt to provide more realistic surface anatomy of the human, these individuals were doing hands-on dissection unencumbered with the earlier medieval anatomical texts and doctrines that were rife with errors. The "typical" surgeon at the beginning of the sixteenth century was nothing more than an unskilled and poorly educated barber surgeon. This surgeon could cut your hair, remove a tooth, and repair a hernia. There were a very few surgeons with either prominent personalities or formal education. The "educated" surgeon having learned medieval dogma remained buried in conjecture and training from centuries of beliefs based on earlier Greco-Roman and later Byzantine teachers and translators who continued to translate and repeated the errors of the past surgical history. In learning and then following these antiquated surgical writings, medieval surgeons often found themselves in conflict with their own bedside observations. As a result of these common conflicts of written text versus what was actually being seen in the anatomical amphitheater, a number of innovative personalities dually learned their surgical material not only as surgeons but also as anatomists. Within the origins of the intellectual climate of the Renaissance, we begin to see profound changes in learning, particularly in the anatomical investigations of the human body.

With the early origins of the Renaissance, we see a renewed interest in human anatomical dissection, anatomical dissections and drawings which at this point had been almost frozen in time for some 1400 years dating to the time of the Alexandrians. From the Byzantine era and through the Middle Ages, anatomy was based on the previous writings of the giants such as Galen of Pergamon who performed their anatomical dissections on non-human subjects and then morphing this information to "human" anatomy. Ironically, it was the Renaissance artist followed by sixteenth-century anatomists and surgeons that led the movement in anatomy away from subservience to the medievalists. With great figures like Leonardo da Vinci (1452–1519), Berengario da Carpi (1470–1550), Johannes Dryander (1500–1560), Andreas Vesalius (1514–1564), and others that was to lead to a new movement based on a hands-on anatomical dissection of the human body. As a result, previous codified anatomical errors, many ensconced since the Greco-Roman era, were to be slowly corrected over the next several centuries. These changes in studies from codified manuscripts to a new and more accurate human anatomy also led to a surge of interest in surgery. The Renaissance surgeon, like the artist, became interested in trying to unravel the intricacies of the human body – without this foundation of knowledge, it would be impossible to correctly treat a disease much less perform a surgical resection. In the area of posterior fossa anatomy and surgery, a number of important Renaissance figures played pivotal roles in bringing forward posterior fossa surgery as both an art and a science.

While neither a surgeon nor a physician, *Leonardo da Vinci* (*1452–1519*) made enormous contributions to both medicine and surgery. Leonardo was the quintessential Renaissance man. Recognized as an artist, an anatomist, and a scientist, Leonardo learned human anatomy, both surface and deep to better provide more realistic artistic creations. Leonardo's anatomical studies were extremely important in providing an early emancipation from the previous medieval teachings. Leonardo's output in anatomical studies led to some 750 separate anatomical drawings. To modern scholars, Leonardo is now considered the founder of iconographic and physiologic anatomy [9–11].

Some of the earliest anatomical drawings on posterior fossa anatomy appear in Leonardo's anatomical studies [9]. To Leonardo we owe the earliest surviving illustrations of the cranial nerves. Figures 1.3 and 1.4, Leonardo did not describe all 12 of the cranial nerves though he was the first to provide some reasonably accurate diagrams. To Leonardo we owe the first illustrations

Fig. 1.3 (**a**) An early copper engraving of Leonardo da Vinci (from the collection of author). (**b**) Leonardo's sketches of the "wax casting" of the ventricles of the brain. In these drawings, the third and fourth ventricles are anatomically outlined for the first time. In the bottom image is an early and rudimentary drawing of the cerebellum and brain stem [9]. (**c**) Da Vinci anatomical illustrations of the ventricular system – enlarged from **b**. There are the earliest known anatomical drawings of the cerebral ventricles – for the first time demonstrating the third ventricle, aqueduct of Sylvius and the fourth ventricle [9]. (**d**) Leonardo's "layered" anatomical studies on the skull, brain, and cranial nerves, what would appear to be the earliest "realistic" anatomical demonstrations of the cranial nerves [9]

of the ventricular system. Using a uniquely designed "wax casting" of the ventricular system, Leonardo was able to detail the anatomical landmarks of these cavities including the fourth ventricle (see Fig. 1.3c). Leonardo's "wax casting" technique was quite innovative and involved removing the brain from the skull and injecting melted wax through the fourth ventricle. Mental tubes were placed in each of the lateral ventricles to allow air to be released. Once the injected wax hardened, the brain was removed leaving behind a wax casting of ventricles.

Fig. 1.4 Leonardo's view of the ventricular system with the cranial nerves detailed at the skull base and exiting to supply the face, mouth, tongue, etc. [9]

In Leonardo's anatomical studies are several investigations that deal with the posterior fossa and its anatomy. His interest in these studies is not clear, as the findings would not have impacted theoretically on his artwork. Leonardo was also not a surgeon so there would not have been any surgical benefit from these studies. Yet his inquisitive mind provides for us some of the earliest and, at the time, the most accurate views of the posterior fossa. Unfortunately, Leonardo's great opus on anatomy, to be published in some 20 volumes, did not appear in print until the twentieth century [9, 10]. Leonardo's anatomical manuscripts did circulate in Italy throughout the sixteenth century among the artistic community; thanks to a close friend and companion, Francesco da Melzi [8]. Leonardo's manuscripts appear to have disappeared from general circulation in the latter half of the sixteenth century, only to be rediscovered in the eighteenth century by William Hunter (1728–1793), a collection that is now part of the Windsor Castle collection, owned by the Queen of England, Elizabeth II. William Hunter was clearly awed by what he saw in Leonardo's drawing and he wrote of his views in a now rare series of eighteenth-century lectures on anatomy – Hunter comments that Leonardo "…was the best anatomist, at that time in the world" [12]. I have included the title and the comment by Hunter on this collection and the anatomy – the first sighting and investigations of these important anatomical illustrations since the sixteenth century. The real mystery is how these important drawings ended up in Scotland in the Windsor Castle collection (Fig. 1.5).

An interesting historical vignette was William Hunter's comment made upon seeing Leonardo's anatomical drawings: (In speaking of Leonardo) "Those very drawings and the writing, are happily found to be preserved in his Majesty's great collection of original drawings. Mr. Dalton, the King's librarian, informed me of this, and at my request procured me the honor of leave to examine them. I expected to see little more than such designs in Anatomy, as might be useful to a painter in his own professions. But I saw, and indeed with astonishment, that Leonardo had been a general and a deep student. When I consider what pains he has taken upon every part of the body, the superiority of his universal genius, his particular excellence in mechanics and hydraulics, and the attention with which such a man would examine and see objects which he was to draw, I am fully persuaded that Leonardo was the best Anatomist, at that time, in the world. We must give the fifteenth century the credit of Leonardo's anatomical studies, as he was 55 years of age at the close of that century" ([12], p 39).

The earliest printed work to appear on neurosurgery, in this case a monograph on head injury, was published in 1518 by *Berengario da Carpi* (*1470–1550*) [13, 14] (see Figs. 1.6 and 1.7). This book was published as a result of Berengario's success in treating a prominent Italian nobleman – Lorenzo dé Medici, Duke of