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ORIGINAL ARTICLE

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The history of spina bifida, hydrocephalus, paraplegia, and incontinence

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Abstract This paper covers the history since ancient times of spina bifida (SB) and its main associated conditions, viz., hydrocephalus, paraplegia and incontinence. Particular stress has been placed on the ancient authors who recognised these conditions. The article concludes with the history of some general aspects of SB and the dilemmas in its treatment.

Keywords History · Spina bifida · Hydrocephalus · Paraplegia · Incontinence

Introduction

The author was the coordinator of the Royal Children's Hospital, Spina Bifida Clinic, Melbourne, Australia, from 1961 to 1978, and this contact with children with SB aroused interest in the history of SB and its associated conditions. Research in the modern period has been particularly helped by the bibliography of Ingraham [48], which lists about 2,300 references from 1556 to 1943. For general background the standard histories of medicine have been used. Evidence for SB goes back to pre-historic times. The literature, considerable since the seventeenth century, swells to a flood in the nineteenth and twentieth centuries and the author has tried to concentrate on its most salient features.

Spina bifida

Spina bifida cystica is obvious at birth and has probably been noticed since time immemorial. Until modern times, however, the chances of survival were slim, and this probably explains the infrequency of reports on the long-term consequences of the disease. Only a small number of skeletons showing SB are dated before. 5000 B.C. One such find was in a cave at Taforalt in Morocco, where about 80 adult and 100 infantile skeletons were discovered. Carbon dating suggests a date of c. B.C.10,000. Some of these skeletons showed splitting of the sacral vertebrae [27]. Abnormalities in the sacrum in Peruvian skeletons dated c. 5000 B.C. have been described [53]. A wide range of diseases and injuries have been noted in skeletons after 5000 B.C., including SB, hydrocephalus, and anencephaly [56, 63, 65]. Pales wrote, "Le spina bifida sacré s'observe à tous les stades et dans toutes les races humaines, en particulier chez les Australiens, les Néo-Calédoniens et les Nègres" [63]. SB is thus of great antiquity.

References in the ancient Greek and Roman writings are uncertain. In the Hippocratic Corpus (c. 500–300 B.C.) in a work not considered to have been written by Hippocrates, there is a description of a fluid-like swelling connected with the spine. It reads: "Another disease takes its rise from the fluid of the head through the vessels in the spinal marrow, from there it makes violence (impulse) in the sacral bone, in which the marrow itself brings (carries) the fluid" [45]. The context would suggest that this is probably not a reference to SB, as Morgagni, who quoted this passage, pointed out [58]. There is no reference to SB in the genuine writings of Hippocrates (c. 460–370 B.C.) and the author was not able to find a reference to it in Celsus (25 B.C.–50 A.D.) or in Galen (130–200 A.D.). The Islamic physicians described hydrocephalus, as is recorded below, but the author has not been able to find a clear reference to SB in their writings. However, this examination has been limited. The literature, written in Arabic, is very large, and only part of it has been translated into Latin or the modern European languages. Gool and Gool refer to a number of excavations from the Middle Ages showing skeletons with sacral malformations [37].

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The first clear description of SB in the West is attributed to Casper Bauhin (1550–1624). Morgagni wrote: "... that the observations of Bauhin were, perhaps, the first that was made upon tumours of this kind" [58]. However, Gool and Gool found a reference not mentioned by Morgagni to a tumour in the neck, probably an occipital or cervical meningocoele, in a work by Pieter Van Foreest (1587) [37] and Ingraham [48] cites a "miracle book" by Boaistuau (1556) as an early reference to SB. The author has not been able to view these two references. Bauhin made some clinical observations in his *Theatrum Anatomicum* (1592) and *Anatomica Historia* (1597), and it is in the former that the comments on SB occur.

SB has been closely connected with the name of Nicholas Tulp (1593–1674) [67, 84, 86], who is familiar to art lovers because he is the central figure in Rembrandt's painting "The Lesson in Anatomy of Dr. Tulp" (1632). Tulp was in charge of anatomy and dissections for the Surgeons' Guild in Amsterdam and was a friend of Rembrandt and prominent in civic life. His best known work is his *Observationes Medicae*, which was first published in 1641 and went through numerous editions. He described a number of patients with SB and included an excellent sketch of the condition, possibly drawn by Rembrandt [88]. Later in the same century, in 1691, Frederick Ruysch (1638–1731) described ten patients. The face of Ruysch is also familiar from Johan Van Neck's painting (1683), "Anatomy of Dr. Frederick Ruysch". In his *Practical Observations in Surgery and Midwifery* he distinguished between the paralytic and non-paralytic forms of the disease and came close to recognising the connection between SB and hydrocephalus [71].

Morgagni (1682–1771), professor at Padua, who in 1760 at the age of 78 published the monumental work that has made him the founder of modern pathological anatomy, clearly recognised that SB can occur with or without hydrocephalus. He wrote: "... that we must not always expect a hydrocephalus to exist in the same patient in whom there is a hydrorachitis", and gave extensive descriptions of both SB and hydrocephalus [58].

Throughout the eighteenth century many monographs, sections of textbooks, and journal articles were written on SB, and nearly 30 are listed in Ingraham's bibliography [48]. However, despite this considerable literature, the disease does not seem to have been widely known, because Underwood (1736–1820), in his *Treatise on the Diseases of Children* (1784) included SB as one of the new diseases recorded in his textbook. The literature from the nineteenth and twentieth centuries is very large, much of it repetitive, and all the varieties of the disease were described. In 1832, Cruveilhier (1791–1874) first advanced the theory of a fault in development as the cause of SB [15], and in 1875, Virchow (1821–1902) described SB occulta [92]. The work of the nineteenth century was consolidated in 1886 in the classic account of the pathological anatomy of the disease by Von Recklinghausen (1833–1910), a masterly description (running to 169 pages) of all forms of the condition [66].

The two main methods of treatment of the sac from the sixteenth century onwards have been ligation/excision and aspiration. Both methods, as would be expected, were disastrous due to sepsis. To try to improve the results, in the middle of the nineteenth century a sclerosing solution of iodine, potassium iodide, and glycerine was injected into the sac, a method popularised by Morton in 1872 [59], though the method had been reported earlier [44, 57]. This method of injecting fluids lingered on until the end of the century, having been recommended by a special committee of the Clinical Society of London that reported its findings in 1885 [13]. Meanwhile, the revolution in surgery introduced by Lister (1827–1912) had occurred and excision of the sac, now made much safer by antiseptic and aseptic techniques, became the method of choice. The operation using musculo-fascial flaps for closure of the sac was introduced by Bayer in 1892 [5] and is substantially the operation still used.

Intensive work on SB at The Royal Children's Hospital (RCH), Melbourne, commenced in 1957 when E. Durham Smith reviewed the patients with SB seen at the hospital from 1943, 306 patients in all [75]. Unexpectedly, 111 of the patients with myelomeningocoele were alive, almost all with significant disabilities. The state of some of the most severely affected children with myelomeningocoele was appalling. It was soon realised that the situation was so complex that only a multidisciplinary team could cope with the problems, and the Spina Bifida Clinic started in April 1961. At this time most of the clinics in Great Britain, Western Europe, the United States, and Canada were small; by far the largest was that of Lorber in Sheffield. The RCH Clinic, the first in Australia, developed rapidly to be one of the largest SB clinics in the world, second only to Lorber's clinic, and at one stage had over 400 children attending.

The three major clinical manifestations of SB-hydrocephalus, paraplegia, and urinary and bowel incontinence – are, like SB itself, easily observable and have been described since ancient times, though not in relationship with SB till the seventeenth century.

Hydrocephalus

Hydrocephalus is one of the most striking abnormalities in infancy. Skulls showing hydrocephalus date back to the late Stone Age. In a grave near Seeburg, about 18 km from Halle, Germany, part of a skull of a 5-to-6-year-old child with hydrocephalus was discovered. It is dated c. 6000 B.C. and is probably the oldest known example of the disease [41]. A number of similar skulls are known from Egyptian burial grounds from archaic to Roman times [21, 56, 63]. As noted above, the Hippocratic Corpus recognised the connection between the cerebro-spinal fluid (CSF) of the brain and the lower part of the body, probably the spinal cord. Herophilus of Alexandria (c. 300 B.C.) knew of the existence of the choroid plexuses. Galen described the meninges, ventricles, and foramen of Munro [32] and the communication between the third and fourth ventricles [31]. Celsus described a swelling of the head, but it is uncertain whether it was a hydrocephalus or a cephalohaematoma [11]. Paul of Aegina (625–690 A.D.), one of the great Byzantine compilers, wrote a book on medicine called *Epitome* (in 7 volumes), which has a section on hydrocephalus with reference to children.

Rhazes (850–923) was the greatest of the Islamic physicians, ranking with Hippocrates and Galen [9]. His most important medical writings are *Al-Hawi or Continens*, a great encyclopaedia, and *Almansoris*, valuable because it quotes from earlier writings that otherwise would have been lost to us. He was the first to devote an entire book to diseases of children, 24 chapters in all, called *De Aegritudinibus Puerorum et Earum Cura*, which was translated into Latin in Venice in 1513. Chapter 3, *De magnitudine capitis puerorum*, is devoted to hydrocephalus. Better known still in the West was Avicenna (980–1037) [9], He was a physician, scientist, poet, and statesman, and his philosophical works place him amongst the greatest of the Islamic philosophers. His *Canon of Medicine* is a vast work in which he tried to codify the whole of medical knowledge, and in it he describes hydrocephalus, though the description is mostly taken from Rhazes. Another mediaeval writer, a contemporary of Avicenna, was Albucasis (or Abulcasis) (936–1013), a Spanish physician and surgeon. He wrote a surgical treatise called *Altasrif*, and chapter 1 describes the surgical treatment of hydrocephalus. In the fifteenth century Metlinger published a book (1473), *Regiment der jungen Kinder*, which has a chapter on hydrocephalus.

There were numerous studies of the brain in the late Middle Ages in Europe, but more important were the works in the sixteenth century, especially that of Andreas Vesalius (1514–1564), the Flemish anatomist and father of modern anatomy. His book *De Humani Corporis Fabrica* is one of the greatest of all medical texts. A book by Brouzet (1754) on hydrocephalus quotes Vesalius: "At Augsburg a little girl of two years has her head so enlarged in less than seven months that I have never seen a man's head to equal it in bulk. Now this affection is what the ancients called hydrocephalus from fluid retained in the head and gradually accumulating. In the case of this little girl this fluid had not collected between the skull and the membrane that covers it externally, nor between it and the skin (where some of the physician's books teach that fluid is sometimes found) but in the cavity of the brain itself, in fact in its right and left ventricle. The cavity and capacity of these had become so much increased and the brain itself so dilated that, – so help me Heaven! – they contained nearly 51/2 pints of fluid" [91]. In the same passage, Vesalius goes on to discuss the thinned membranous condition of the skull and the wide separation of the sutures. He mentions the normal findings in the basal part and cerebellum and remarks that the child retained her senses till the day of her death and had no convulsions.

In the sixteenth century Thomas Phayre (Phaire) (1510–1560), contemporary with Vesalius, in his book *The Boke of Children* (1545), the first paediatric text written in English, describes hydrocephalus. In the same century in 1565 Simon de Vallambert wrote the first paediatric text in French, *Cinq Livres, de la Maniere de Nourir et Gouverner les Infants des leur Naissance*, which contains a section on hydrocephalus. He wrote: "They do not all die young. Today at Chastellerauff, where I now live, there is a girl, daughter of an apothecary, who has an enormous enlargement of the head; she is seven or eight years old and eats well, but she has very small thighs and legs and cannot hold herself upright" [83]. In the seventeenth century there were extensive monographs on paediatrics and in the same century Richard Lower explored the formation and circulation of the CSF and the mechanism of hydrocephalus. Astruc (1684–1766), a French physician, is most famous for his works on sexual subjects, *De morbis veneris libri sex*, but he also wrote a book (English edition, 1746) describing the site of the fluid in hydrocephalus, stating that the fluid was "betwixt the dura and pia mater, or in the convolutions of the brain, or in the ventricles, which last is the most frequent of all" [83].

The pressure of the CSF had long been recognised, but the circulation had been imperfectly understood. Modern study commenced with Magendie between 1827 and 1842, summarised in his book (1842) *Recherges philosophiques et cliniques sur le liquide cephalbrachien ou cerebrospina*. He described the medial foramen in the fourth ventricle named after him. Faivre proved that the choroid plexuses produced the fluid [26] and Luschka wrote extensively on the nervous system in *Die Structur der serösen Haute des Menschen* (1851); he also discovered the lateral foramina named after him. These workers were followed by Key and Retzius, who described the circulation in detail in *Studier nervosystmets anatomi* (1872), and these studies were confirmed and extended by Dandy [17–19]. At the end of the nineteenth century the Arnold-Chiari phenomenon in the mechanism of hydrocephalus was described. This was originally described by Arnold in 1894 [3] and by Chiari in 1895 [12] and was recognised as an important mechanism in the hydrocephalus of SB [49].

The treatment of hydrocephalus proved to be difficult. The ancient Greeks used incision and drainage. Paul of Aegina advised against surgical interference, but he commented that some physicians used trephining. The Islamic physicians used cautery with a hot iron on the suture lines and temporal veins. These measures were often accompanied by dehydration using purges and diuretics and the use of compression bandages to the head, and these methods continued well into the nineteenth century. Continuous drainage to the outside was used from the 1880s onwards. All of these measures were ineffective, and in the pre-Lister era, disastrous because of sepsis. As in the treatment of SB, injection of iodine into the ventricles was tried to destroy the epithelial lining, and in the early twentieth century destruction of the choroid plexus was attempted by radiation, cauterisation, or excision.

The failure of external drainage turned surgeons in the 1890s to internal drainage, and many attempts were made to drain the fluid into the sub-dural spaces by tubes or by making a channel with linen threads [20]. The best known operation using this concept is that of Torkildsen (1939) [87]. It was more logical to transfer the fluid to the blood stream – the basis of the modern operations – and this commenced with Payr in 1908, who inserted a tube from the ventricle to the longitudinal sinus, jugular vein, or common facial vein [64]. His attempt failed because of inadequate shunt materials and valves. In 1909 McClure used a transplanted vein as the shunt channel, transplanting the fluid into the jugular vein, but this also was not successful [54]. The modern shunt tubes date from the work of Nulsen and Spitz of Philadelphia and John Holter, an engineer, published in 1952 [62]. Since then, a number of valve designs have been used and the material itself has been improved. Scanning and imaging techniques have also greatly facilitated diagnosis and treatment. With all their problems, the shunts have transformed the outlook for children with SB and hydrocephalus.

Paraplegia

Paraplegia has been studied since ancient times, but not in relation to SB. However, the studies on paraplegia from injuries have provided the basic knowledge of the spinal cord that has been essential to understanding the paraplegia of SB.

Head and spinal injuries are shown in many Stone Age skeletons, and in the Edwin Smith Surgical Papyrus, discovered at Luxor in 1862, are clear descriptions of the resulting clinical state. This papyrus dates from the seventeenth century B.C., when it was copied by a scribe from earlier material, and much of the original material dates from the 3rd millennium B.C. Forty-eight patients are described, including 27 with head trauma and 6 with spinal trauma. Most interesting is case 31, which gives an excellent account of quadriplegia and incontinence of urine and sexual function from spinal injury. "Title – Instructions concerning a dislocation in a vertebra of (his) neck. Examination – If thou examanist a man having a dislocation in a vertebra of his neck, shouldest thou find him unconscious of his two arms (and) his two legs on account of it, while his phallus is erected on account of it, (and) urine drops from his member without his knowing it, his flesh has received wind, his two eyes are blood-shot; it is a dislocation of his neck extending to his backbone, which causes him to be unconscious of his two arms (and) his two legs. If, however, the middle vertebra of his neck is dislocated, it is an emmissio seminis which befalls his phallus. An ailment not to be treated" [94].

Hippocrates also dealt with spinal injuries and described the results of backward and forward dislocations of the vertebrae [46]. Herophilus of Chalcedon (c. 300 B.C.) distinguished between motor and sensory nerves and also described the ventricles of the brain. In a work quoted by Rufus of Ephesus (early second century A.D.), Herophilus wrote: "Amongst the nerves that issue from the brain and spinal marrow the motor and sensory are called voluntary and tensor" [70]. Hippocrates knew that damage to one side of the brain produced spasms on the other side of the body [47], and Aretaeus the Cappodocian (second and third century A.D.) elaborated on this phenomenon [1, 34]. Celsus noted that injury to the lower parts of the spinal cord produced paralysis of the legs and urinary retention or incontinence [11].

The brilliant experiments of Galen clearly established the motor and sensory functions of the spinal cord. Galen was by far the greatest physician after Hippocrates, and his influence continued until the sixteenth century. There was much confusion in the Greek writings between nerves, ligaments, and tendons, and these tissues were distinguished by Galen [29], who gave a detailed account of dissection of the spinal cord. He used living apes and monkeys for his experiments and described the results of sectioning the cord at various levels [30, 33]. This knowledge was clearly accepted and reaffirmed by Caelius Aurelianus in the fifth century A.D. [22]. Paraplegia was well known in the Middle Ages and the healing of the paralytic, the lame, and the blind is a common theme in mosaics, an example being the twelfth-century mosaic in the cathedral of Monreale, Sicily.

Vesalius commenced more detailed studies and dissection of the spinal cord. He wrote: "Even so if anyone may have considered examining the function of the dorsal medulla, it will be seen when the medulla has been injured how the parts below the injury lose sensation and motion." [91]. These studies were elaborated in the eighteenth century by Francois Pourfour du Petit (1664–1741) and in the nineteenth century by Ludwig Turck (1810–1868), and especially by Paul Flechsig (1847–1929). Although the spinal cord had been dissected, little attempt had been made to study the internal structure of the cord. This was commenced by Pourfour de Petit, followed by Johann Jacob Huber (1707–1778), who gave the first accurate detailed description of the cord in *De medulla spinale speciatimde nervis ab ea provenientibus commentatis cum adjunctis iconibus* (1741). Huber's work was extended by the studies of FJlix Vicq d'Azyr (1748–1794) in *Traité d'anatomie et de physiologie* (1786); he paid much more attention to the division of the cord into columns, originally suggested by Huber.

The above two workers studied the cord with the naked eye. In the seventeenth century the microscopic appearance of the cord was described by Leeuwenhoek in a letter of 1677 [51]. Benedict Stilling (1810–1879) developed microscopic techniques for examining the cord by serial sections and was able to give much more detail in *Neue Untersuchungen über dem Bau des Rückenmarks*. Hotop (1859), and he was followed by A. von Koelliker (1817–1905) in studies from 1841 to 1896. Koelliker used fixer stains and was able to give more consideration to the grey and white matter of the cord and the origin and distribution of the motor and sensory fibres. In classic experiments, Charles Bell (1774–1842) studied the functions of the motor roots [6], and finally, Francois Magendie (1783–1855) gave definite experimental proof in *Recherges philosophiques et cliniques sur le liquide cephalorachien ou cerebrospinal* (1842) of the functions of both the motor and sensory roots. These detailed studies were confirmed and extended by Johann Muller (1801–1858), Stilling, and Charles Edouard Brown-Séquard (1817–1894).

Once the functions of the motor and sensory roots had been established, the distribution of these roots began to be studied. Conrad Eckhard (1822–1905) started studies on the motor (myotomes) and sensory (dermatomes) distributions [23]. These studies were extended by Lüdwig Turck, Charles Sherrington, Head and Campbell using herpes zoster [40], and Foerster using the effects of root trauma or surgery [28]. With the increasing survival of patients with SB in the 1960s, renewed studies were made of the segmental distribution of the spinal cord by Sharrard [73], Durham Smith [75], Stark and Baker [82], Menelaus [55], Stark [81], and others. It became obvious that the problem was more complicated than previously postulated.

Meanwhile, functional studies were made of the cord itself, revolving around the concept of reflex action. Detailed study of reflexes did not start until the seventeenth century. Ancient writers had noticed the distinction between voluntary and involuntary movement as described by Aristotle [2] and Galen [31]. There was speculation on reflex action in the seventeenth century by Descartes and Thomas Willis and more secure work in the eighteenth century by Robert Whytt and Galvani, but the beginning of the study of reflex action commences with Marshall Hall (1790–1857). He clearly established the reflex as a fundamental feature of nervous action. He introduced the term 'reflex' and showed that it could be distinguished from other types of movement and that it produced what today would be called "muscle tone" [38]. In other experiments using salamanders, frogs, and turtles, he divided the spinal cord between the upper and lower extremities and noted the normal movements and sensation above the level of the lesion [38].

In the nineteenth century the neurone theory and the anatomical basis of the spinal reflexes were established. The central connections of the spinal cord were at last known, and in 1891 Waldeyer-Hartz could describe the various components of the spinal reflexes [93]. The clinical significance of reflexes was established by Wilhelm Heinrick Erb (1840–1921) [25] and Babinski (1857–1932) [4]. The whole field of neurones, synapses, and reflexes was integrated by Sherrington in *The Integrative Action of the Nervous System* (1906). During the period 1870–1900, the main methods of clinical neurological examination were developed by many workers, and these and other facts have been incorporated in the modern neurological examination. Here is all the essential material forming the basis of the neurological diagnosis of the type of paralysis and level of the lesion in SB.

The treatment of paraplegia, whether due to injury, SB, or other spinal diseases, has been depressing. As described above, the Edwin Smith Surgical Papyrus of the 2nd millennium B.C. stated that it was "an ailment not to be treated", and this was the standard approach until well into the twentieth century. The decisive change in the outlook came in 1944 through the pioneering work of Ludwig Guttmann at the Stoke Mandeville Hospital near Oxford. This work has transformed the rehabilitation of people with paraplegia.

The major orthopaedic problems relate to the hips and spine. Over the years orthopaedic treatment has become more targeted, resulting in a marked reduction in the duration of hospitalisation and disruption of the child's education and social life. Splints have been used since ancient times; the earliest that have come to light have come from an Egyptian tomb of the 5th Dynasty, c. 2400 B.C. [76]. Hippocrates used splints extensively in his instructions on fractures and dislocations. Fabricus (1560–1634) described many types of splints and braces, and they have been in frequent use since the seventeenth century. The splints used in SB were at first complicated and heavy, but the introduction of plastic splints in the mid-1970s made them much lighter and more socially acceptable. Most children who are going to be functional walkers, either without splints or with minimal splinting, do so by the time they enter primary school. For children who were not functional walkers, extensive splints were previously used in the school period, but were soon abandoned because the child was much more mobile in a wheelchair and more able to take part in school and recreational activities.

Incontinence

Incontinence has also been recognised from ancient times. Attention was drawn earlier to the urinary symptoms in spinal injury in the Edwin Smith Egyptian papyrus [94] and in the writings of Hippocrates [46]. Incontinence is also mentioned in the Egyptian Ebers Papyrus, a medical papyrus dated c. 1550 B.C. and listing over 1,000 medical recipes; for incontinence the papyrus recommends juniper berries, cypress and beer, and opiates if the child cried! The most extensive investigations were, however, carried out by Galen in his experiments involving section of the spinal cord. He showed that in diseases of the spinal cord there was atony of the bladder and also demonstrated in animal experiments the valvular action of the bladder neck [33].

Aetius (502–575), born in Mesopotamia (now Iraq), was eminent enough to be physician at the Byzantine court at Constantinople and was the first Christian physician of note. He wrote a huge treatise called *Tetrabiblion* in which are included 26 chapters on the care of children and their diseases. Incontinence in children was differentiated as either continual and dribbling or occasional and nocturnal, and the former was attributed to paralysis of the sphincter of the bladder. He was the first author to make an extensive study of the urine and gave a detailed description of its appearance in relation to diagnosis and treatment.

Following Aetius, the Islamic physicians made detailed studies of the urine and described many of the urinary diseases, and Avicenna wrote specifically on incontinence. In the fifteenth century Paolo Bagellardo wrote a treatise on diseases of children, *Libellus de Aegritudinibus Infantum*. It was printed in Padua and was one the first printed medical books. Part 2 of 22 chapters has a chapter on "incontinence of urine". In the next century, Sebastian Ostricher produced a scholarly paediatric text; this contained a chapter on incontinence entitled *De meientibus*. In the seventeenth century (1684) Stephen Blankaart published a book in Dutch on diseases of children. It contains a chapter on "bed-wetting" and also deals with faecal incontinence. Lorenz Heister (1683–1758) also dealt with incontinence. The above are general references to incontinence; Hildanus' *Observationum et curationum chirurgicarum centuria tertia* (1614) seems to have been the first work to link incontinence directly with SB. In the eighteenth century Morgagni described most of the urinary diseases that are known today.

For males, penile bags, and for both sexes, urinary pads and intermittent catheters have been used for SB since the seventeenth century. Catheters, mainly of bronze or lead, are very ancient, having been used in ancient Egypt and in the Hippocratic era for obstruction [61]. Fabricus (1537–1619) was the first to use nonmetallic catheters. In the nineteenth century rubber and silk made an excellent combination and the twentieth century saw the introduction of various types of plastic catheters. The results of these methods were poor. The renal failure, smell, skin excoriation, and social stigma were the incentive to use diversion of the urine as an alternative.

From 1852 attempts were made to divert the urine. In that year, John Simon at St. Thomas Hospital in London transplanted the ureters into the bowel for exstrophy of the bladder [74]. However, because of the bowel incontinence, this method was useless for the incontinence of SB and so other solutions were explored, especially bringing the ureter to the skin surface. Cutaneous ureterostomy was first suggested by Gigon in 1856 [35] and carried out experimentally and in humans for a variety of conditions by Gluck and Zeller in 1881 [36], Edmund and Balance in 1886 [24], Le Dentu in 1890 [50], and Reginald Harrison in 1897 [39]. Finally, in 1906 Thorkild Rovsing brought through enough ureter to form a nipple and applied a small silver cup [69].

Problems of stenosis and leakage of urine made this procedure unsatisfactory so that surgeons turned to another solution, viz., transplanting the ureters into a loop of ileum and using this to form a nipple – the principle of the ileal conduit operation. In 1888 Tizzoni and Poggi of Bologna performed this experimentally [85], and in 1911 Zaaijer [95] and in 1935 Seiffert [72] performed it in humans. Bricker became the main exponent, and his techniques described in 1950 are the basis of the modern operation [7].

In the 1980s there was a reversion of the treatment of urinary incontinence from the ileal conduit to the use of intermittent catheterisation and urinary pads. With silicone protectives, better urinary antiseptics, and much more efficient, odour-free pads, catheterisation is now considered to be "more natural" and more socially acceptable by the patients and parents. This method has been assisted by the use of artificial sphincters for the bladder and by augmented cystoplasty to increase the capacity of the bladder and lower the urinary pressure. The use of the ileal conduit clearly showed a better prognosis for renal failure than the old methods; whether the new methods will prove as effective remains to be proved. A new technique is, however, now available for ultimate renal failure – the renal transplant.

Discussion

Some general aspects of SB can only be mentioned briefly: detailed discussion of these diverse fields is beyond the scope of this paper. The general rehabilitation of disabled children has advanced markedly over the last few decades. The longer life-span focussed attention on the education of children with SB, the reaction of the children and their parents to the disability, the sexual potentialities of the adults, on the employment of adults with SB, and the integration of children and adults into the community. This produced a steady stream of studies from the late 1960s onwards. The increased life-span also made possible an intensive study of the quality of life of the survivors [77]. The interest has not only been on quality of life for its own sake, but also because attempts have been made to correlate the findings with those in new-born children, and so give a factual basis for the difficult decisions that have to be made in the neonatal period [52, 78].

From the 1970 s increased use has been made of the rapidly growing knowledge of pre-natal diagnosis. In 1972 Brock and Sutcliffe described the pre-natal presence of alpha-fetoprotein in the amniotic fluid in open cases of SB [8], and later authors confirmed the presence of this substance in the amniotic fluid and in maternal serum and its use in the diagnosis of SB [89, 90]. The accuracy of pre-natal diagnosis has been further enhanced by ultrasonography. Pre-natal diagnosis, and abortion if SB is diagnosed, has greatly reduced the number of live-born infants with SB. However, a survey of congenital abnormalities in Victoria, Australia, from 1987 to 1994 inclusive, showed that there was no decline in the total number of anencephalic and SB fetuses [14].

Abortion as a means of prevention is not an ideal method, but the search for the cause of the disorder, and so effective prevention, is proving elusive. The most promising method of prevention is nutritional. In 1964 and 1965 the possibility of folic acid preventing SB was suggested [42, 43]. In the 1980s, by using vitamin supplements before conception and in the first trimester, there appeared to be a fall in the incidence of SB [79, 80]. Further studies showed that folic acid was the essential element in these supplements and that its effect on the incidence of SB was significant [16, 60, 68]. Folic acid supplementation is now recommended to all women contemplating pregnancy, especially those who have had a previous child with a neural-tube defect. It is expected that the absolute number of fetuses with anencephaly or SB will fall as this procedure becomes more widespread.

The ethical problems raised in the treatment of patients with SB, especially infants, continue to occupy the profession. Infanticide has been firmly rejected. Infanticide for children with abnormalities (and for other social reasons) was widespread in the ancient world, has been common throughout the centuries, and is still present in some countries. It was strongly opposed by the Christian Church. The Christian Emperor Valentinian in 374 A.D. made infanticide a capital offence, and this was incorporated into subsequent legal codes and has been absorbed into the Western legal systems. While rejecting infanticide, doctors for centuries have been reluctant to give active treatment to children with severe abnormalities. Until the twentieth century, the results with SB were so poor and, in its myelomeningocoele form, its associated conditions so disabling, that active treatment was generally withheld, and this is still the practice in many countries racked by war, displacement, poverty, and starvation. With the more effective treatment of SB and its associated conditions in the second half of the twentieth century, active treatment at birth or soon thereafter is now practicable. In some cases where the infant has gross abnormalities (often with other abnormalities in addition to SB) active treatment is deferred until survival is assured [52, 78].

The decision between immediate active treatment and deferred treatment remains difficult in some children, but most surgeons hesitate to submit an infant to operations and extensive procedures when death in infancy is almost certain with or without active treatment. The philosophic background to the decision to defer or withhold treatment in certain patients is the distinction between "ordinary" (obligatory) care, such as feeding, simple nursing, and relief of pain, and "extraordinary" (optional) care, such as extensive medical procedures, life-support systems, etc. This distinction, itself controversial, dates from the Middle Ages and the Spanish Dominican monk Domingo Báñez (sixteenth century) is credited with introducing these terms. Since that time the distinction has been extensively discussed, especially in books on moral theology, and over the last 50 years, there has been even more discussion in professional journals and the media.

The distinction is not an absolute one; what is defined as "extraordinary" varies according to the time, place, circumstances, and the sophistication and resources of medical services. This philosophy is expressed in the ironic poem of the nineteenth century poet, Arthur Hugh Clough,

> "Thou shalt not kill; but need'st not strive
> Officiously to keep alive"

and more formally in the official *Catechism of the Catholic Church* (1994) [10]. The distinction has been widely used by moralists, the church, individual physicians, and medical associations. The principles have changed little down the centuries; what is new today is the expertise available to keep people alive.

SB and its associated disorders have a very long history, and connected with this history have been some of the greatest names in the history of medicine. The achievements down the centuries bring the study of the disease into perspective. Inevitably, we must work in the light shed by our predecessors, and all we can do is to turn up that light and peer a little further into the darkness of the unknown.

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