Patient Perspectives

18

Graham Flint

Contents

18.1 Introduction

The preceding chapters in this book have dealt with many different technical aspects of the subject of syringomyelia and its related conditions, ranging from what causes them to develop in the first place through to how they are best treated, given the current state of medical knowledge. At the end of the day, however, there is an individual who is suffering from the effects of the disease. That person will suffer all the normal human emotions that go with uncertainty about a condition with a strange name and about which most of the medical profession seem to know relatively little. It is important that those health-care professionals who deal with these conditions have at least some appreciation of what the patient goes through, in terms of both physical and emotional distress. Many of these experiences may not be obvious to the doctor, working in the clinical environment and with limited consultation time.

Below four of the authors give brief summaries of their own experiences as patients. Each has also spoken to many other patients with syringomyelia and Chiari. Their own case histories can therefore be put into context by a review of the experiences of other patients. Together these teach us a good deal about the many needs of people who find themselves having to live with these rather frightening conditions.

With Sue Line, Susan Kember, Rebecca Dodwell-Pitt and Lynn Burton

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18.2 Case Histories

18.2.1 Personal Story 1

"*On December 31st 1970, having enjoyed a meal with a friend, we both decided to visit a colleague. It was a very cold night, black ice was forecast and we certainly found some! I had been travelling at 30 mph so was able to control some of the skid but, with mud on the road under the ice, the car hit a small bank, spun and landed in the middle of the road, leaving me on the verge and my passenger in a ditch. My passenger had a dislocated collar bone and concussion. I had a severe spinal injury involving T5 and T6, in addition to broken ribs and damage to my right arm. I spent the following five and half months in a spinal injuries unit. I was left a complete paraplegic with sensation only in the upper torso.*

"*After a further 4 weeks at home I returned to my employer albeit in a modified capacity. Fate was still not on my side, however and, many years later, on my way to a meeting one October evening in 1987, a car went into the side of mine causing me to have a whiplash injury. I contacted a solicitor about claiming back the excess payment on my insurance. His enquiry "have you noticed any problems since the incident?" prompted a chain of events that I could not have envisaged.*

"*By then I had already noticed that, to drive, I needed to wear leather palmed gloves. At home and at work I was beginning to drop things more frequently. I had occasional but intense pain at the base of my skull. Sometimes it was difficult to concentrate. Sometimes even light touch on the base of my neck produced great discomfort. I was also having some problems distinguishing temperatures. I was not sure what the problem was. Were these symptoms the result of the most recent car accident or something else?*

"*On the advice of my solicitor I returned to my spinal injuries unit. My strongest recollection of that time was of multiple visits, with repeated examinations, including testing by pin prick, which leaves you feeling like a pin cushion, even if you can only feel some of the jabs. I remember being in a state of disbelief when the diagnosis of syringomyelia was made. Wasn't it enough that I was coping with the spinal injury and had got my life back together, without having to deal with this latest revelation?*

"*Next I was sent to see a neurosurgeon. My sister accompanied me to the appointment and I was really glad she did. The vivid description of what he intended to do was quite frightening as he proposed to "sever the spinal cord" to reduce the possibility of further syrinx forming and causing more problems. Thinking about the consequences and the possible detrimental effects upon my life left me dazed. I don't think either my sister or I will forget the drive home. Lots of questions came into my head about the surgery. What if it meant I could no longer tell when I wanted to go to the toilet? How would I cope being in hospital for a period of time? My mum and my sister were my carers and, although I could still do lots for myself, what would happen after this operation on my spine? Did I really want this operation? How long would I have to wait before surgery if I did decide to go ahead?*

"*While waiting to go into hospital, I set about reading up about the condition. At this time most of the information available was for professionals and very scary for the lay person to read. There was a lot of technical jargon and words which I didn't understand. I remembered the support group leaflet that the neurosurgeon had given me and at last plucked up the courage to speak to someone. Talking to somebody who had undergone surgery helped me realise that perhaps the future was not as bleak as I imagined. I then tried to carry on as normal but the prospect of surgery was always at the back of my mind. It is not an easy thing to face and the hope that there is an alternative around the corner keeps you searching.*

"*Plans had, nevertheless, been put in place for my surgery and, come the admission day, I was still very apprehensive whether this was the right decision but deep down I suspected it was. By chance there was a meeting of the patient support group that week and I found it really helpful to meet other people with the condition and all its variants. My problems seemed insignificant compared to what some individuals had to bear. Yet the people there showed camaraderie and*

supported each other and the charity. It was a real eye opening experience. I plucked up the courage to ask my surgeon about the consequences of the operation, especially with regard to bladder and bowel function. I was very proud that, despite having no feeling on the surface of my body, I could still tell when I wanted to go to the toilet and I was very anxious that I might lose this after the surgery.

"Tuesday, and the big day had arrived. The porters came to take me to theatre and I started laughing. In fact I didn't stop until the anaesthetic took hold. My next memory is being on the intensive care unit with oxygen pipes in my nostrils and feeling very thirsty. The noise level was high and the lights very bright. Most of the time I was drifting in and out of sleep. I had a urinary catheter which had been my big nightmare. After 2 days I returned to the ward and one thing I remember vividly was an increase in body spasm. This was like being in a vice, holding me rigid and I felt like I had to fight for every breath. A bout could last anything from a few minutes to more than an hour. Increasing my anti-spasm medication did not help. My memory of the next three post-operative weeks is sketchy. The physiotherapist guided me through the process of becoming active again and regaining confidence in my ability to resume my activities. Turning to avoid pressure sores was very painful and was something I won't forget! Gritting your teeth also helped when the dressings were changed. After a few days I started the process of retraining my bladder. My goal was to rid myself of the urinary catheter as soon as possible. Three weeks after the operation I went home and started the process of getting back to work.

"The first benefit that I noticed was the ability to "walk" my thumb to every finger, one at a time in a sequence. This was a fantastic feeling and confirmed I had made the right decision as I had something tangible to compare with my pre-operative state. Many years on I can look back and say there have been several benefits. Initially it was great not to drop items so frequently and this was one of the first things family and friends noticed. My confidence increased and I was more willing to try new things and situations. Would I have the operation again? I hope I don't have to but yes, I would, as the benefits have outweighed the initial concerns and worries.

"It is difficult to know whether my remaining symptoms are the result of syringomyelia, the original spinal injury or something else. Alternatively, as I am now a senior citizen, are they simply age related? Recently I have found that it is becoming more difficult to hold as much in my hands as I used to. Picking up small objects was difficult before but is now more so. Feeling objects and temperature awareness have become more of a problem. I have developed severe sleep apnoea and have to using a Continuous Positive Air Pressure (CPAP) pump at night. High blood pressure and elevated cholesterol are two other conditions that have become part of my life. These are controlled with medication and regular checks. I can't say that I notice I have them. I have acupuncture, approximately every 6 weeks, which is helping to keep me well. It helps my balance, circulation, general well-being and also my spasm, which means I can maintain a lower dosage of medication.

"The support of family and friends has been crucial in encouraging me to carry on when sometimes I am in great discomfort. Not knowing which of the conditions is the trigger to my symptoms is very frustrating and makes it harder to find a solution. I like to think I don't live with my conditions; rather my conditions live with me. I count myself as a survivor and not a sufferer. It is important that I work in partnership with all the health care professionals concerned with my well-being. I see myself at the hub of all this activity but sometimes I don't know what to do to get the answers to some of my questions. There is a wealth of information available if you know where and how to look and, importantly, can understand it. A source of understandable information, that is not frightening, is essential for patients, family and friends. There is only a small percentage of people with a spinal injury who also have syringomyelia so linking up and supporting one another helps, even if that is only possible sporadically.

"An on-going concern of mine is how the condition will change my ability to do things in the

future. I feel like a pioneer as I don't know what's coming next. At the moment I just keep doing what seems to help me cope and I take 1 day at a time. I feel there is a need for more research into the correlation between the consequence of spinal cord injury, syringomyelia and decompression surgery and whether other symptoms, such as sleep apnoea and high blood pressure, are connected. The Ann Conroy Trust has gone a long way to helping individuals tell their stories, both in the magazine and on their web site together with providing support and information from professionals. Even though my condition was diagnosed and dealt with all those years ago, I am still learning from the information available, and will continue to do so."

Sue Line

18.2.2 Personal Story 2

"*In 1994, after suffering significant headaches for some time, I visited my GP who referred me to a consultant physician who in turn carried out several tests, before sending me for an MR scan. This revealed that I had a Chiari I malformation. I was then referred to a neurologist and subsequently to a neurosurgeon. The neurosurgeon explained what a Chiari malformation was but felt that in my case it was not significant enough to warrant surgery. I and my family were of course concerned but took comfort in the fact that there was no immediate need for treatment. My life continued as normal but my headaches progressively worsened. I reached the point where I was unable to put my head forward and laughing, sneezing or coughing would start a major headache which could last for several hours. I was also experiencing some frightening episodes where I felt as though I was fading away. Altogether, life was becoming pretty miserable.*

"*I was referred once again to a neurologist who, after another MRI, explained why I was having these symptoms and suggested that I should see the neurosurgeon again. At this time there was absolutely no information available about my condition other than terrifying internet sites. I felt very alone. I had also decided that I needed to find a neurosurgeon with particular experience in carrying out any surgery which was necessary. This I did, with help from my brother. After consultation with the specialist I agreed to proceed to surgery. Indeed, by now I was anxious to have the surgery as soon as possible.*

"*The operation was successful and my subsequent recovery proceeded very well. Prior to surgery I had been put in touch with a syringomyelia specialist nurse and she had been able to give me some idea of how my life would be when I came out of hospital. The surgery was very beneficial in eliminating my headaches and my life soon returned to normal. I was now able to put my head down without any concern. With each month I became stronger and 6 months after my operation I was well enough to undergo further surgery for an unrelated condition. Despite this I went on to experience some severe pain in my lower back. I underwent a full spine MRI which revealed that I had a small syringomyelia cavity in the lower part of my spinal cord. This had not been evident in my original scan which had been confined to the head and upper part of my spine. However the discomfort subsided and, 11 years on, I have hardly any symptoms from my Chiari or syringomyelia.*

"*At the time of my diagnosis there was little information on my condition and hardly any support. With hindsight and the information that I now have, I feel I was misadvised by my original surgeon. After my operation I was invited by my neurosurgeon to join and help a charity group, now known as the Ann Conroy Trust, which was involved with helping patients with Chiari malformation and syringomyelia. This led to both my husband and I becoming increasingly involved with the running of this organisation. In addition to its other activities, the Charity provides medically approved information on these conditions, for patients and their families and carers. I assisted running the helpline for newly diagnosed sufferers, as well as those who had already undergone surgery. I would have been extremely grateful if any of these services had been available to me when I was a newly diagnosed patient.*

"*My hopes for the future would be that a more comprehensive* support *system could be set up for patients and that that there would be more nurses trained, available and prepared to speak and give practical advice to sufferers, particularly those facing surgery. I would like to see improved opportunities for the education of neurosurgeons in the management of syringomyelia and Chiari. I would like information to be more readily available to family doctors and non-specialists, so that they might be less dismissive of symptoms that they cannot immediately diagnose. Most of all I would like to see more coordinated research into these conditions and the publication of results in a more structured manner. This is why I worked for many years as a volunteer with the Ann Conroy Trust, to try and facilitate such changes.*"

Susan Kember

18.2.3 Personal Story 3

"*I first noticed that something was not right when, on a couple of occasions, I experienced tingling and numbness on the left side of my face and in my left hand. I thought that I must be tired, working late and rushing around. I then fell down the stairs at home a couple of times. There was then an episode of numbness in my chest and arms so I went to hospital where the doctor thought it might have been a panic attack. I next went to see my family doctor and he thought it might be epilepsy. As I work as a nurse, I spoke to one of my consultants who requested an MRI scan. When the results came through he sat me down and reassured me that it was not anything sinister, which was a huge relief as I work on a cancer unit. He explained a little about Chiari malformation but then referred me to a local neurosurgeon. I informed my family doctor about developments and he commented that I was probably the only patient he would see in his professional lifetime with this condition.*

"*When I saw the neurosurgeon he explained that not everyone needs surgery and that sometimes symptoms remain stable and individuals can live with them. He suggested that we should try and manage my condition conservatively for the time being. Unfortunately, with time, my arm tingling became worse and I was starting to suffer severe headaches, which gave me a bit of a 'short fuse,' and I would get frustrated easily. I also began to experience panic attacks, during which I was convinced that something dreadful was going to happen.*

"*Many months later, having decided that I now needed surgery, I went into hospital for the operation. I was woken early on the day of the procedure. I was meant to go to theatre in the morning but an emergency delayed things so it was the afternoon when I went down. My nerves, by then, were getting the better of me; much longer and I'd have changed my mind. I spoke to my neurosurgeon and the anaesthetist in the anaesthetic room and the next thing I remember is the neurosurgeon coming into the recovery area. I tried to sit up as I thought we were still having the same conversation and did not realise straight away that I'd had the surgery. My head certainly hurt though and an injection of codeine helped for an hour at the most. Paracetamol didn't help at all. The next morning I was the first to be moved to the ordinary ward. Three days after surgery I felt well enough to go home.*

"*At home I tried to do too much. That night I started vomiting and this continued for 24 h. I had visitors but I couldn't keep awake to see them. I was readmitted to hospital in the early hours. I had a CT scan to check for hydrocephalus and then a lumbar puncture. The next day another patient told me that she had needed three shunts for her condition and I was frightened that the same might happen to me. My neurosurgeon then told me that my test results were fine and I could go home again. I spent the next 3 months recovering properly. Unfortunately I started getting the headaches back just before I returned to work and I began to worry that this would be the case for the rest of my life. I decided that, if this was so, I simply couldn't live with it and I actually contemplated suicide. I then returned to hospital for another lumbar puncture and, generally speaking since then, my residual symptoms have been minimal and manageable.*

"*I was well informed about the surgery itself although I was so desperate to have the operation done and get rid of the headaches that I didn't really listen to talk about the risks of surgery. It wasn't until I spoke to my neurosurgeon about 3 years ago that I could fully appreciate what could have happened if things had gone wrong. At the time I felt that I had enough information available to me, if I chose to access it. I had the contact number of the specialist nurse and the neurosurgeon's secretary. I had also been given a newsletter from a patient self-help group but I didn't read this until after my surgery. When I did, it was quite depressing and not very helpful. It mentioned people who had recently died and contained recipes and articles that were not relevant but no information about the condition. There was information on the internet but this was mainly from overseas sites and showed lots of gruesome pictures. I didn't choose to access any patient support group prior to my surgery but relied on the couple of visits to the neurosurgeon and specialist nurse, along with support from my family and friends, who were all very supportive during the time of the operation and thereafter.*

"*Nine years on, I am certain that I made the right decision. I had been keen to get the surgery over and done with and to move on with my life. The Chiari had already dominated 2 years of my life. I still have some residual symptoms such as irritability, occasional headaches, neck stiffness and poor balance but I can now smile to myself, particularly when people asked if they can 'pick my brains' about something, or when I refer to my condition as having been 'always at the back of my mind'.*"

Rebecca Dodwell-Pitt

18.2.4 Personal Story 4

"*Looking back to when I was in my early 20s I had what I thought at the time was a one off horrendous migraine. At the time I was working as a singer and the 'migraine' followed a new dance routine, which included movements in which I bent my head forwards. The migraine left me shaky for a couple of days but I soon forgot about it, and forgot the dance routine too.*

"*I first experienced any real problems at the age of 33, 3 days after the birth of my daughter. She was in a Moses basket and I leant over to lift her and my head suddenly pounded and I couldn't move. I sat down and the pounding subsided. During the night I found it impossible to lift my head from the pillow and the only way I could get up was to roll out of bed. My GP called the morning after, to give the new baby a quick check over. He thought that maybe it was cervical spondylosis aggravated by the delivery. Happily, everything settled down over the next couple of weeks. I next gave birth to a son, in 1989, with no postnatal problems.*

"*Over the next few years I had different symptoms; tingling in my face and around my mouth, tingling in my fingers, blurred vision and a feeling that my spine pushed into my neck whenever I walked. When I visited my doctor the symptoms were put down to just one of those unexplained things. In due course I stopped reporting them.*

"*In 1998 I was out for the evening with friends and noticed that every time I laughed my head pounded with pain, I found myself holding the back of my neck every time. I visited my doctor and cervical spondylosis was again mentioned. Once more X-rays were unremarkable. This time, however, the symptoms didn't go away. The pounding headaches occurred almost daily, along with the tingling in my arms and fingers, I started to experience balance problems and blurred vision.*

"*In 1999 I was referred to rheumatologist, who diagnosed fibromyalgia. Over the next 2 years the symptoms got worse. Rheumatoid arthritis was then suspected and I was treated with steroids. Things didn't improve and in 2001 I was referred to a neurologist who thought I might have multiple sclerosis. I was again treated with steroids, as well as being given amitriptyline and tramadol for pain relief. As the symptoms were getting worse I was admitted to hospital for a lumbar puncture and an MRI scan. The good news was, 'there were no plaques on the brain' to suggest multiple sclerosis. The bad news was 'the*

base of my brain had come down into what should be a space above the spinal cord' but as I didn't seem to have any symptoms I could go home. I was speechless. The symptoms I was experiencing were the reason for the MRI.

"By March 2004 my neurologist seemed to have decided that I must be neurotic and all my symptoms were psychological. In the May I duly received an appointment to see a psychiatrist. This was a low point for me and for the first time I wept, mainly because I felt that I had wasted the time and money of my GP, who had always been there for me and had never doubted my symptoms were genuine. I saw him and started to apologise for wasting his time. He assured me I had not and would refer me to a multiple sclerosis specialist in London. I did, however, also see the psychiatrist and was told that I was not neurotic and that my symptoms were indeed neurological. That was a great relief but still left me no closer to a diagnosis. When I then saw the consultant in London he did not want to discuss MS but was, instead, extremely concerned about the base of my brain; 'it's jammed well down into the spinal cord with no room for CSF flow' He promised to discuss this with colleagues and get back to me. I'm still waiting.

"A friend then e-mailed me, to ask if I had a diagnosis. She knew someone with the same symptoms and gave me the name Arnold-Chiari malformation type 1. She told me to go to my GP with it. Of course I Googled it first and was confronted with the words 'a serious and complicated condition of the base of the brain and spinal cord which, if left untreated, can lead to total and permanent paralysis . I rang my GP immediately and he asked me to print out the article and get to the surgery. By the time I got there he had printed articles from the BMJ which, of course, were far less sensational. Although very scary, it was a relief, after 6 long years, to have, finally, a diagnosis. It was around this time that I found the Ann Conroy Trust, which proved to be an invaluable support during what was a frightening time for me.

"Within 2 days I was seeing a neurosurgeon, who said I needed a craniovertebral decompression. He explained that the surgery was major and had risks but if I was willing to have the operation he would perform it. I duly had my decompression surgery, on the 1st September 2005. I woke after surgery in the intensive care unit. It was dark, thunder was banging and lightening was sparking across the sky when a man with a goatee beard leaned over me, to ask how my pain was. For a fleeting moment I thought things had gone wrong and I had been sent down to you-know-where! Of course he was a nurse, asking if I needed any more pain relief. I was in hospital for 2 weeks. On discharge I had a temperature and was vomiting but it was considered that I would still be alright to go home. Thank goodness for my own doctor, who prescribed antibiotics and other strong medication.

"It soon became apparent that my symptoms hadn't resolved and, after 11 months, I saw a different neurologist who referred me to another neurosurgeon. He explained the risks of operating in the same area again so we agreed to wait and see how things went. A year later symptoms were getting worse and the pounding in the head was happening several times a day, even lifting my head when turning over during the night was enough to start the pounding. The tingling in my arms and hands was worse, as was my balance. Another MR scan revealed that scar tissue was now blocking CSF flow posteriorly and causing Valsalva-type headaches.

"Eventually I underwent revisional surgery, in February 2007. This was not without complications, including some damage to a small but important artery, which was embedded in scar tissue. I became unwell and was found to have meningitis. I was in hospital for 4 weeks.

"I now still have ongoing problems. Postoperative scar tissue is again pulling my spinal cord backwards into the area of decompression, once again blocking posterior CSF flow, although there is some space for CSF flow anteriorly. I had a number of lumbar punctures performed and these improved many of my symptoms so, eventually, I had a lumboperitoneal shunt fitted. This helped for 3 weeks but then over draining caused new problems and, 3 weeks later, the shunt was closed. I am now scheduled for surgery to have a gravity assist valve fitted to the shunt.

"*Arnold Chiari 1 malformation has without a doubt changed my life. I am limited in what I am able to do. I tire easily. I can't walk far and have to use a walking stick for balance and am housebound most of the time. I can no longer do many things, such as walking in the Lake District with my husband. I was a mature student at university in 1999 but the symptoms became so bad I had to end my studies. I nevertheless maintain a positive outlook. I have become chairman of the Ann Conroy Trust and help to answer calls to the Charity's helpline, as I know only too well what a scary, lonely place the world is, after the diagnosis of Chiari malformation.*"

Lynn Burton

18.3 Learning from Patients

These accounts illustrate clearly the experiences and emotions that accompanied four different patient journeys (Table 18.1). After the initial onset of symptoms, any individual will normally wait a while, anticipating that they will settle down before very long. When they continue, a visit is made to the family doctor who will certainly not make an initial diagnosis of syringomyelia or Chiari. When symptoms persist a referral will likely follow, to a hospital specialist, for further assessment. Not infrequently this leads to MR imaging and the diagnosis is made. Sometimes, however, diagnoses such as fibromyalgia may delay the true cause being revealed. The patient may then have to endure worsening symptoms, which fail to respond to various treatment regimes. The delay in the correct diagnosis being made is a common source of complaint from both syringomyelia and Chiari patients, but this is, of course, to be expected, with such uncommon ailments.

Table 18.1 Common experiences

Worry and uncertainty prior to diagnosis
Delays in diagnosis
Relief and new worries when diagnosis is made
Alarm about proposed brain surgery
Value of talking to other patients
Dependence upon family and friends for support
Value of patient information material
Positive and negative impact of websites and support groups
The need to have confidence in the neurosurgeon
Fear of possible complications of surgery
Post-operative discomforts and the experience of ITU care
Relief when symptoms improve
Concerns when symptoms return
Coming to terms with residual symptoms
A wish to help fellow sufferers

With the eventual diagnosis comes a combination of relief and further anxieties. Numerous new questions arise and answers are not always forthcoming. It is at this stage that the hospital specialist has a vital role to play, in gaining the patients confidence and trust. Unless and until the individual is adequately informed about their condition, he or she will not be able to make a rational decision about the most appropriate course of treatment to follow. Surgical intervention for uncomplicated Chiari is seldom essential and even the presence of an associated syrinx does not necessarily make an operation mandatory. The patient needs time to come to terms with matters, with careful explanation and guidance from the specialist. If an operation is the chosen route then the surgeon must give clear information about the procedure.

A frequent concern amongst patients and carers is the lack of general support available to them. Often, before deciding to submit to surgery, the patient may look to a fellow sufferer for some counsel. An increase in social networking sites and mobile communication technology has meant that patients can do this online. Small local support groups may also be set up, and these certainly have a role to play, provided that they are well run and do not become a forum for propagating complaints and negative attitudes. Running a support group is, however, not an easy task. Quite apart from the time commitment and the need to be available as often as possible, the volunteer has to develop counselling skills, often without the benefit of any formal training. For the caller, however, the opportunity to talk, at last, to somebody who seems to understand what they are going through, is a great relief.

Table 18.2 Common concerns

Gaining access to specialists with sufficient expertise
Adequate time for concerns to be heard by clinicians
To be believed and not treated as being neurotic
Knowing which symptoms do and which do not arise from the condition
Lack of readily available information, for patient, about the conditions
Lack of educational material for family practitioners
Access to professional, informed counselling
Fear of being isolated
Fear of progression to severe disability or death
Special needs of children as patients

Different callers have individual needs, of course, but there a number of common themes (Table 18.2). Gaining access to specialists with sufficient expertise and experience in treating syringomyelia and Chiari is a common concern amongst patients. Many will be seen initially by their local neurology or neurosurgery service, and not all units will have an individual with a special interest in managing these conditions. The initial consultation may very well leave the patient confused and frightened, particularly if the prospect of brain or spinal cord surgery is raised at the outset. Patients commonly resort to the Internet to gain some further understanding about these conditions with the strange-sounding names. All too often this "research" adds to the patient's fears, raising the spectre of progressive, painful deterioration, to a state of physical helplessness. Patients, therefore, should be guided by their own professional health-care advisors and use information from other sources simply as a means of better understanding what their medical advisors say to them. The patient desperately needs to be listened to and to know that the specialist understands something about the problem and what the patient is going through. The patient wants to know which symptoms are likely to be caused by the Chiari or the syrinx and which are not. Is there a risk of coming to serious harm? What other treatment options are available? He or she then needs time to consider matters and to read an intelligible, lay account which does not overdramatise matters. The patient will probably then come back with further questions and may well ask if there is a fellow sufferer to whom he or she can talk. Above all the patient wants to be believed and not left feeling that the doctors regard him or her as suffering primarily from stress.

Another common complaint amongst patients is that there is also a lack of information available for health-care professionals who are not specialists in this field. Most patients report that their family doctor has little or no awareness of syringomyelia and Chiari, but this, of course, is largely due to the low number of patients that they will see in their professional lifetime. Some patients ask if charities could distribute information to all family practices, but, clearly, these would probably just sit on a shelf, and if a patient did present with syringomyelia or Chiari, the leaflet would probably have been misplaced by then or simply be overlooked. A more effective answer would be the provision of Web-based information for non-specialist medical practitioners. It is probably not helpful for patients to visit their doctor with a large amount of information downloaded from the Internet as the doctor will almost certainly not find time to read it.

When it comes to surgery, as a proposed treatment, the patient must be told, in a clear but sympathetic way, just what is involved and what are the likely benefits and also what could go wrong, as well as what might happen if the condition were left untreated. Volunteer counsellors receive, from time to time, calls from people who have not done well after surgery. They say that they would not have undergone the operation had they known what was going to happen. Lay advisors cannot, of course, give opinions on matters of medical detail. They will certainly not suggest whether or not the caller should undergo surgery. The patient must always question the surgeon before any operation and be quite satisfied about the risks as well as the benefits of the procedure. These may prove to be, quite literally, vital issues. There will, of course, be other, less critical questions but which are, nevertheless, important for the patient to have answered (Table 18.3).

Even after a successful operation it is unlikely that the patient will be able to put matters to rest

Table 18.3 Frequently asked questions

Where will I have the surgery?
How long will I be in hospital?
How long will I be in the operating theatre?
What size wound will I have and what stitches will I have?
How much hair will I lose?
When can I wash my hair?
How long will in need to take off work/college?
What is the total recovery period?
Will I be able to fly?
Will I be able to drive?
What follow-up will I receive?
Will my headaches/other pains improve?
Will my hindbrain hernia ever return?
Will my syringomyelia resolve?
Could my syringomyelia refill at a later date?
What happens if I have problems in the future?

completely. Persistence of some symptoms and recurrence of others will generate fears that the condition may recur. Anticipation of further pain and discomfort can sometimes be as distressing as the symptoms themselves. Here, once again, patient support groups can play a useful role. Surgeons vary in their approach to management at this stage, some simply stating that there is no more role for surgery. Others become more involved in exploring alternative avenues for providing the patient with some relief from their continuing suffering. There are several support groups and sources of information worldwide, and some of the more well-established groups are listed in Chap. 22.

Finally, many patients express concern about the apparent lack of any research being carried out into syringomyelia and Chiari. It is clear that a better coordinated approach to research, internationally, is needed.

Further Reading

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