BORN UNTO TROUBLE*

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We describe unexpected adverse psychological and social results in three adult men with severe Little's disease who were made more mobile by successful surgery.

It is well known that ill-judged surgical intervention in patients with Little's disease may make their limbs straighter but decrease their mobility, particularly if operation is not followed by adequate rehabilitation. It is less well recognised that surgery, successful in improving a patient's mobility and independence, may be considered 'psychologically' disastrous by the patient or the onlooker.

The three patients to be described were in a residential home for adults of both sexes suffering from Little's disease. They were seen at the home by a consultant in Physical Medicine who referred them for an orthopaedic opinion.

CASE REPORTS

Case 1
This man was thirty-two years of age at the time of operation on his knees.

Social background—The patient was a foundling, thought to have been born in 1923. He lived in an orphanage for the first two years of life and was then found to be physically handicapped and with a speech defect, so he was admitted to a long-term hospital for the mentally subnormal. While there, he had operations in each adductor region and stabilisation of the left tarsus. He received no formal education. When the patient was aged about thirty-one years the hospital almoner, learning of the foundation of the Spastics Society in 1954, was able to get him admitted to one of their residential centres.

Condition on referral—His speech was very limited and extremely difficult to understand. He claimed to be able to walk a mile (this was unverified). There was severe spastic tetraplegia with moderate atrophy, the right arm being least affected. Walking produced a severe dynamic flexion deformity of the knees (Fig. 1) so that he progressed with the knees flexed at about a right angle, though fixed flexion was only about 20 degrees in the left knee and 10 degrees in the right. There were associated dynamic flexion deforms of the hips and equinus of the ankles, but the main spasm was in the knee flexors. There was a 90 degrees fixed flexion contracture of the left wrist.

At conference there was some doubt about the advisability of embarking upon operation but it was agreed to do a posterior release upon each knee and not to treat the left wrist.

The knees were operated upon at an interval of four weeks. This resulted in marked improvement in stance. On walking, the knees and hips were only slightly flexed and the feet were plantigrade (Fig. 2).

The complication—At the Centre the patient had been taught to weave, and within a year became expert and a perfectionist at the loom, winning prizes at local handicraft exhibitions, county shows, and even in national competitions. He was invited to appear personally at local events to demonstrate his skill, and became something of a minor celebrity in the locality, exciting much comment upon the grotesqueness of his physique.

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* "Man is born unto trouble as the sparks fly upward." The Book of Job, chapter 5, verse 7.
radiance of his smile and the skill and perfection of his handiwork.

Although unable to read or write, he could produce complicated geometric designs from memory and his order book was filled with requests for his original patterns. His customers visited him at the Centre and would congratulate him, expressing amazement at his dexterity despite the deformity of arms and legs. He enjoyed these visits greatly, and his knees would jerk into further flexion spasms with excitement and pleasure. This was possibly the fullest period of his life; he had come to terms with his handicap, his industrious nature had found a satisfying occupation and he had achieved the attention and esteem that had, up till now, been lacking in his life.

Although he had complied eagerly with the suggestion that he should have operations to enable him to walk more easily, he found the surgical procedure more traumatic than he had expected. In spite of his handicap he was unused to, and unable to bear, pain with any stoicism. He wept frequently and could not be comforted. Upon his discharge from hospital he attended physiotherapy sessions only with reluctance, and his rehabilitation took almost two years. When at last he was able to walk with a stick, he seemed less than pleased and proud of his new mobility. His appearance, while less grotesque, excited less comment and admiration. He became morose, for he could not adjust to the changes that had taken place.

Though deserving of even greater sympathy because of the painful and psychological adjustments needed, he found that he got rather less than compassionate treatment from his peers, “making too much fuss” and “not grateful” being the general consensus of opinion at the Centre. To visitors the perfection of his weaving seemed less of an achievement by one with only a slight limp than had previously been the case, and their commendations were less frequent and less vocal. He had lost more than he had gained and gradually isolated himself, withdrawing into his shell. After some years of increasing disharmony he asked to be transferred to another institution, where he has been seen recently. In his present home he is the only resident with a significant speech defect and he has been able to use this to regain the sympathy which was withdrawn from him when his leg deformities were improved.

Case 2
This man was aged twenty-five to thirty years at the time of operation.

Social background—The baby was deserted by his mother in his first year, and though his father tried to care for him, his work entailed moving around the country and the child was admitted for short-term care to many different institutions. He had no schooling and never learned to read or write. The documentary evidence as to his age is in conflict, but at about the age of eleven to sixteen he was admitted for long-term care to an institution for mentally subnormal patients. While there he was seen by the local medical officer of health, who thought he had been mistakenly placed and arranged for the boy to be transferred to a Cheshire Home. However, the Home applied for his discharge on the grounds that they were no longer able to contain him because of his stealing and dirty habits and he was transferred back to the institution for mentally subnormal patients. After a few more months he was admitted to the residential home for spastics. He lived there for six to seven years and his growing desire to overcome his inability to walk and to be like normal people led to his referral, at the age of twenty-five to thirty years, for consideration for surgery.

Condition on referral—There was severe spasticity of both legs and the right arm. The left arm was a little affected. He had never walked. With support, he could stand on the left leg (Fig. 3) but spent most of the days with his hips and knees flexed (Fig. 4). The hips showed flexion and adduction spasms. There were sweat rashes in the flexures. Extension of the right knee was limited by 90 degrees. The patient was warned that failure of surgery might necessitate amputation of the right leg.

Operations—The right knee was arthrodesed: the bones were shortened by about 5 centimetres and fixation was provided by an intramedullary nail, Charnley clamps and a plaster-of-Paris cylinder. Despite some infection and several operations for drainage of abscesses, the arthrodesis was solid within six months, and the Künschert nail was removed. Three months after that the left knee was straightened by a posterior release operation, followed by a turnbuckle plaster. A year from the beginning of treatment he could walk a few yards but was severely hampered by flexion spasms of the hips. After a further six months of walking practice he had a left obturator neurectomy with division of the adductors, the psas and the anterior part of the gluteus medius. Three months later the flexors of the right hip were divided.

Progress—After three and a half years of treatment the patient, who had never even been able to stand unaided, could sit with comfort, get in and out of a chair alone and walk about the Institution with no other help than the walking aid (Fig. 5). He had a 5 centimetres raise to the right shoe.

The complication—Although this patient was of broadly normal intelligence, the grievous physical handicap, his aggressive and uncontrolled manner and the deprivations of his early life had produced personality problems of a formidable nature. He had undergone the lengthy and painful surgical procedures with great stoicism, buoyed up by the hope and expectation that, after completion, he would walk out of residential care. Although he achieved the ability to walk with an aid when attending physiotherapy sessions for a few months, he would not practise at the Centre and, after a while, refused to attend for further walking training. He refused to attend physiotherapy sessions, becoming, upon his return to the Centre,
Case 2. Figure 3—A 25-year-old patient with severe spasticity in the lower limbs and right upper limb. He is shown trying to stand. Figure 4—His usual sitting position. Figure 5—The right knee has been arthrodesed and hip flexors divided; on the left lower limb the procedures were: posterior release of knee, obturator neurectomy and division of adductors, ilipsoas and gluteus medius. He could now sit properly and walk well using an aid.

totally demanding—unwilling and perhaps unable to do anything at all, and certainly not to walk or even to stand.

Protracted social work-counselling revealed that his anger and frustration stemmed from the fact that he had hoped and expected that the operations would not only straighten his limbs but would remove his handicaps entirely, and in some miraculous way transform him into a successful man, tall and able to command respect, attract the kind of woman he admired, give him a job involving freedom and money and, in addition, the material status-symbols of car and dress that he had long desired. Suggestions that if he were able to make an effort and work at it increased mobility might lead eventually to sheltered employment and life in a hostel, were totally rejected, as was the notion that he might form a relationship with one of the spastic girls at the Centre.

During the next eighteen months, he wove a fantasy about himself in which he was the victim of surgical errors that had turned him from a normal young man into a spastic invalid, and spoke darkly of "experiments" to which he had been an unwilling guinea-pig, "mucking me about" as he put it. Staff wearily refuted the tales he told to shocked visitors and reproached him for his irresponsibility—to no avail. The depth of his disappointment and his sense of grievance were too strong to allow reason to prevail. He was found a place in a hostel in another part of the country, where nearby sheltered workshop facilities offered a chance for him to make a new and more independent life, were he willing to forgo the personal attentions he had hitherto demanded. He left the Centre with high hopes, determined to succeed, and "to show them".

Unfortunately, his inability to achieve any independence precluded the hope of succeeding in a hostel where the staff ratio was lower and where cooperation and the will for a less dependent way of life was essential for success. He returned to the original centre more than four times, having been found unsuitable for hostel life. After many incidents, he was placed in a small adult house unit, designed to provide a bridging between those institutions offering total care and the hostels. When seen again recently, he appeared dirty and unkempt in spite of the provision of dentures and a wig. He has become engaged to be married to another of the residents at the house unit and intends to get a flat in London, marry, and live independently. It seems doubtful whether this will come about—for the couple both need personal help. The patient does not walk or stand at all: "not worth it", he says.

Case 3
This man was aged eighteen years at the time of operation. Social background—He was the first child of his mother's
third marriage, with seven siblings, and attended a Catholic school for the physically handicapped. At the age of seventeen years, as a school leaver, he was admitted to the Spastics Society Home.

He had many unrealistic adolescent fantasies and, in particular, the desire for a car. His attitude was that “something must be done”.

Progress—For a time after the conclusion of these leg operations he suffered from “touch hunger”, missing the frequent handling he had formerly required, and because of this he pretended that he was unable to feed himself.

Four years from the start of treatment he could walk well across the room with a walking aid and could manage on buses and trains unaccompanied. His gait was grossly lurching without an aid and he was advised to continue with its use, though he wanted to use only sticks (Fig. 7).

The complication—He had progressed well after the operations and was delighted with his new-found mobility and freedom, but he refused to accept the responsibilities attached to it and expected to go on receiving total physical care, though this was no longer necessary. He became an impossible companion for the other residents and the Warden was forced to ask for the patient’s transfer. He also was admitted to a hostel with outwork at a sheltered workshop nearby, but he refused to take his share of the simple domestic duties expected of him, such as washing up, and after only one month he was discharged back to the Spastics Centre. More recently he has been transferred to a centre with full-time educational facilities but his expectations remain unrealistic.

He intended sitting “O” level examinations in mathematics and sociology, “writing” the examinations
by dictation or typewriter; he was unsure which. He has applied for an invalid motor car and has recently been tested and promised a road trial with an electric carriage ("electric chair" might be a more appropriate term) before being provided with a petrol-driven machine.

DISCUSSION

In three cases the surgeon expected the patients and their companions to be pleased, if not actually grateful, for the improvement in mobility resulting from their treatment. But in the first case an obvious source of sympathy had been removed and the patient felt far more deprived than benefited, and even his weaving at the loom suffered as a result.

In the second case the patient, who in fact was illiterate, had a quite unrealistic fantasy about what would happen when he could walk and he was resentful that the operations had failed to make him tall and attractive, and to command a highly paid job. Both of these patients became dissatisfied with their former environment, moved to wider pastures at their own wish, and then found that the grass there was not so green after all.

The third patient is delighted with his newly-found independence. It is his companions who are distressed, as they find he has grasped at this independence without accepting the social obligations that go with it.

A detailed description of the social and psychological aspects of these patients is being published elsewhere.

These patients had certain things in common. All had spent their formative years in institutions. They all had a very deprived family background and no normal education. None of them actively sought treatment. "The System" offered treatment to them and they willingly complied with the suggestion. They were all physically if not mentally adult and so relatively fixed in their attitudes and expectations. The surgeon was quite unaware that, by lessening a physical disability, he might be removing a source of comforting sympathy (Case 1) or, by only partly fulfilling a fantasy which could not be wholly achieved, the agony of failure was so much the worse to the patient after all his boastings, the discomfort of multiple operations, and months of rehabilitation (Case 2).

The last patient thinks he is cured and if his ambition does not quite stretch to the moon, at least he feels it should embrace driving on the motorways. While sympathising with those who criticise the mechanical imperfections of the invalid three-wheeled vehicle, should we not consider the possible psychological inadequacy of some of the disabled drivers that may be let loose upon the roads? This can be just as important as their physical infirmity and could contribute to the high accident rate of these vehicles.

There is an important difference between the congenitally handicapped and those with acquired disabilities. The latter have known normality.

In congenitally handicapped adults there is an established body image and it must not be forgotten that to them the handicap is normality. It may not be sufficient to tell the patient of the "improvement" he may gain from surgery without understanding the patient's expectations and his notion of independence and normality. Therefore, it is very important to find out what the patient means by independence and, if need be, seek the help of a trained counsellor. The feelings of loss and grief experienced by a "normal" person who acquires a disability from disease or accident are well recognised and understood. The relevant support is usually given, together with the sympathy and help needed towards rehabilitation. On the other hand, if the congenitally handicapped have been able to turn their disability to some advantage in obtaining extra sympathy or bodily care, they too may feel loss and grief and fear for the future after undergoing corrective surgery, but they may not be given the relevant support. The emotional shock they suffer may not be recognised, or the patient may think it unworthy to grieve, and fear to be thought ungrateful if he does.

New-found mobility for those who have previously never walked and may have been in institutions all of their lives, can induce great trauma. The patient may become contemptuous of his former peers and impatient to leave his former home. Yet he may not be fit for the outside world, craving freedom and independence without experience of responsibility.

Failure to recognise such symptoms may impede rehabilitation and the patient may find himself unable to achieve what were previously simple goals. He may have "touch hunger", missing being physically handled, as in Case 3.

CONCLUSION

We now realise that many congenitally handicapped persons have had limited opportunity to achieve discernment and may be unrealistic, so it behoves us to define more accurately what the patient means by cure and help. We should be more cautious in offering, and embarking upon, a line of treatment unsought by the patient, and, if it is undertaken, we should be more searching in the social as well as the physical follow-up.

A problem remains. Clearly, patients in long-stay institutions should be allowed the benefit of active treatment if it is available. On the other hand, we, as doctors or workers in the social sphere, have a special responsibility to be careful lest, by offering the patient a small physical improvement, we upset adversely the delicate symbiosis he has come to accept with his disability.

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