A Survey of Mentally Subnormal Patients with Orthopaedic Abnormalities at Muckamore Abbey Hospital

W. V. James, F.R.C.S.
Withers Orthopaedic Centre, Musgrave Park Hospital, and the Ulster Hospital, Belfast

In 1967 it was decided that an orthopaedic surgeon should pay regular visits to Muckamore Abbey. It was soon realised that instead of seeing selected cases it would be more productive to carry out a survey of all the patients. This would mean that every patient would be examined at least once by an orthopaedic surgeon. It so happens that at Muckamore Abbey the patients with orthopaedic problems tend to be concentrated in some of the villas, thus by the time the first 300 patients had been seen, the majority of those with orthopaedic abnormalities had been found. By the time that 400 patients had been examined, a considerable number of orthopaedic problems had accumulated and those patients that had undergone surgery were returning for follow-up. The survey was stopped after seeing 450 patients, and this communication describes the main findings.

Patients with Orthopaedic Abnormalities

The object of the survey was to discover those patients that would benefit by orthopaedic treatment. Whilst the primary diagnosis was of interest from the prognostic point of view, it was the functional state of the patient that was of prime interest. Many had purely orthopaedic conditions such as valgus feet and knock knees, but this did not materially affect their functional capacity. Disorders of muscle control however were far more common and greatly affected the capacity to be independent.

Of the 450 patients, 299 were found to have conditions of orthopaedic interest.

The Primary Diagnosis

The primary diagnoses are shown in Table I. The term amentia is not very precise and in it there were a group that had evidence of mental and physical deterioration. The latter revealed itself by muscle wasting.

Of the sixty-five mongols, thirty-three were sixteen or under. Only seven were unable to walk. It is of interest to note that almost a third of them had a

| Table I |
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| **Primary Diagnoses** |
| Primary Amentia and Microcephaly 341 | Gargoylism .... .... .... 1 |
| Mongol .... .... .... 65 | Incontinence pigmenti .... .... 1 |
| Hydrocephaly .... .... .... 18 | Marfan's syndrome .... .... 1 |
| Phenylketonuria .... .... .... 7 | Cretin .... .... .... 1 |
| Birth Injury .... .... .... 4 | *Cri du chat* .... .... .... 1 |
| Tuberose sclerosis .... .... .... 3 | Glycine deficiency .... .... .... 1 |
| Tuberculous meningitis .... .... .... 2 | Selenium poisoning .... .... .... 1 |
| Hereditary ataxia .... .... .... 2 | Encephalitis .... .... .... 1 |
valgus deformity of the feet, which is probably associated with the ligamentous laxity. This was slightly commoner than the wide cleft between the big and second toes. Three mongols had congenital skeletal abnormalities, two having hemivertebrae, and one had phocomelia.

Of the patients with hydrocephaly, ten were confined to bed by a combination of mental retardation and paralysis. Six could walk, and two were confined to a chair due to lower limb paralysis.

**Disorders of Muscle Control**

There were 139 patients with disorders of muscle control, the majority being children. Of these 113 had mainly increased muscle tone, 19 decreased muscle tone, five athetosis, and two were ataxic. These types of motor disturbance tended to be combined in most patients. The relationship of the disorder to the functional capacity is noted in Table II.

|                  | Increased tone | Decreased tone | Athetosis | Ataxia |
|------------------|----------------|----------------|-----------|--------|
| Bedridden        | 33             | 13             |           |        |
| Chairbound       | 24             | 5              | 2         |        |
| Walked a few steps | 12             |                | 2*        |        |
| Walked reasonably | 33             | 1              | 3         |        |
| Walked normally  | 11             |                |           |        |

* one has subsequently died

**Degree of Mental Subnormality**

This varied from slight educational subnormality with an I.Q. of 70 to complete lack of comprehension of the surroundings. Where it was possible, the intelligence quotient had been estimated, but there were great difficulties with those who had severe muscle abnormalities or who were blind or deaf. It was found that the intelligence quotient was of little help in deciding whether or not a child would have the mental capacity to make use of any improvement that was made in the physical state. It was found more useful to assess roughly the degree of motivation of the child. Unfortunately there is no available method of expressing this quantitatively. The motivation had to be estimated by the “spark in the eye” or whether the child showed physically a desire to move around. Often the patient had a desire to move but was limited by physical incapacity, and this resulted in frustration tantrums. This may possibly give a quantitative measurement of motivation. Motivation also gave rise to difficulties in assessing the results of operative treatment, for motivation may vary and sometimes many months went by before a child suddenly decided to walk and make use of an operative physical improvement.

**The Bedridden Patients**

Broadly speaking the severity of the physical state corresponded to the mental state. Forty-seven bedridden patients were seen of whom twenty-nine were male and eighteen were female. Fourteen were five and under, twenty-three between
six and sixteen, seven were from seventeen to forty, and three were over forty. The primary diagnosis in thirty-seven was amentia, and hydrocephaly in ten. Six were blind, deaf, or unable to speak. Twelve of the forty-seven had flexion contractures of the knees, and eight had contractures of the hips, six of the hips being dislocated. It must be realised that in nearly all of these patients the gross mental abnormalities alone would have confined them to bed.

The children who were confined to bed with quadriplegia in flexion had the most severe contractures. It appeared that if the knees and hips were flexed, they also fell to one side so that one hip was flexed and abducted, and the other hip flexed and adducted. Of eight patients with a flexed and adducted hip, six of these hips were dislocated. It must be noted that the nursing at Muckamore Abbey is of the highest quality, and the staff constantly straighten the limbs, yet in spite of this contractures occurred. It must also be noted that all these patients were incontinent and helpless, yet not a single bed sore was seen. Those who were completely helpless also developed gravity deformities of the skull and thorax.

The Chairbound Patients

Thirty-nine patients were chairbound, of whom eighteen were male and twenty-one female. One was under five, nineteen were six to sixteen years of age, thirteen were between seventeen and forty, and six were over forty. Eight had normal tone, twenty-three had increased muscle tone, six had decreased muscle tone, and two had athetosis.

Most were confined to chair life because of physical incapacity, but some had no motivation to walk. It was noted that of the thirty-nine, eleven had developed a scoliosis, which in some cases had become severe. It was noted that these were all children with rather poor muscle tone, and slumped in their chairs. It was also noted that some of the children who had previously been classified as bedridden, could be re-classified as chairbound when fitted with a thoracic support. The nursing staff noted that these children appeared to be far more contented when they could sit up with the aid of a support, and could see the world around them.

Those who could Walk a Few Steps

There were fourteen patients who could walk a few steps, nine male and five female. Ten were under sixteen years and four over. All showed increased muscle tone, but two also had marked athetosis and two had ataxia. These patients could walk in a walking machine, but did not have enough control to use crutches, they did however show a desire to walk.

Those who Walked Reasonably Well

There were forty-three patients in this group, twenty male and twenty-three female. Three were age five years and under, twenty between six and sixteen, eleven between seventeen and forty, and nine over forty. All except one could be classified as having cerebral palsy, and all had reasonable motivation. They walked with difficulty due to adduction of the hips and flexion deformities of the hips and knees, and plantar-flexion of the feet. It was in this group that the greatest number of operative procedures were undertaken in an effort to reduce the spasticity and deformity of the limbs.
TREATMENT

Non-operative

Physiotherapy is still the primary form of treatment for these patients. The treatment is aimed at encouraging the children to use what capacity they have, rather than the correction of individual deformities. It is of little use the physiotherapist ensuring that a muscle is used in a certain way for thirty minutes daily if during the rest of the day the lesson is lost. To encourage a child to walk demands an intimate knowledge of the psychology of that particular child, and the treatment must be individual. Many of the children are unco-operative to a direct approach. Adequate apparatus is available at Muckamore Abbey, but with such individual attention necessary, the limitations of treatment lay in the staff available. It is true to say, however, that the treatment that is given is of the highest standard. The nursing staff also spent a large part of their time encouraging the patients to make use of their physical capacity.

Ten children were fitted with supportive corsets. The majority of these were of leather, and had a neck brace. An attempt was made to use Plastizote, which is easier technically to produce, but the support was in no way superior to that made from leather. Eight children were supplied with calipers. These were only used when absolutely necessary, for the very weight of the caliper was a hindrance. Several children had their calipers removed as it was considered that in fact they were hindered by them, and it must be recorded that some of the parents objected.

Operative Treatment

In all, eighty-one orthopaedic procedures were performed. Four of these were for fractures of the neck of the femur, and two for syndactyly. The remainder were performed for the correction of muscle imbalance and deformity.

Of the eighty-one operations, sixty-three were performed on forty-one children for the correction of lower limb deformities and muscle imbalance. It must be realised that a child may have several operations, but in cerebral palsy they are performed at intervals, so that their individual effects can be noted.

The Problems Arising from the Survey

There were three main groups of patients that required investigation and help.

1. The Bedridden Children with Flexion Deformities

It would seem that helpless children who are confined to bed, and who have an increase in flexor tone of their muscles, will eventually develop contractures in spite of good nursing and physiotherapy. Contractures are not desirable, for joint movements become painful to the child, and the variety of positions in which the child can lie becomes very restricted. This means that their nursing is painful and difficult, and they become liable to gravity deformities and possibly serious complications. The nursing staff found dressing and undressing of the patient difficult, and there had to be considerable care over the prevention of bed sores.

In established contractures, there is a great reluctance to perform operative correction upon these children, especially if they are markedly mentally retarded. One of the major complications of the contractures is dislocation of the hip. To perform an open reduction and to release contractures is a major procedure, the benefits of which are not obvious to some when the child is seen with the lower limbs neatly covered by bedclothes.

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2. Those who are unable to sit correctly without aid

Many children had weak spinal muscles and were unable to sit up without aid. It was noted that those who were already sitting tended to develop a scoliosis, which in time would give rise to respiratory problems. These children were fitted with a leather supporting brace. Many also had weakness of the neck muscles, and were fitted with a Milwaukee brace to support the head. To fit a leather support is a lengthy and expensive procedure, and some experiments were carried out with Plastizote supports. The patients appeared to be far more comfortable when in their supports, especially those with weak neck muscles who appreciated that they were able to see far more around them. The support also controlled or prevented the development of scoliosis. Some of the bedridden children are now able to sit up, and could be re-classified as chairbound. The nursing staff and parents also appreciated the provision of a support. Many of the children were difficult to lift or move due to their lack of muscle tone, but when wearing the support they could easily be lifted.

3. Those who had difficulty in walking

Orthopaedic procedures on mentally normal children who suffer from disorders of muscle tone can produce reasonably predictable results. With mentally subnormal children the results are difficult to predict and are often disappointing. There is great difficulty in judging whether or not the child is going to make use of any physical improvement. Many procedures used in cerebral palsy involve the re-education of muscles, but, even if the muscles learn their new roles, the child must have the desire to make full use of the new conditions. It has already been mentioned that there is no quantitative method of assessing the motivation of a child. A decision on the suitability of a child for operation can only be arrived at by a long period of observation.

There are further pitfalls in operative treatment in that some of the children show cyclical variations in activity, and it may be many months before a child will show any improvement, or, alternatively, a child who has been active, may decide not to walk for a time.

Conclusion

As a result of the survey, some forty-nine patients have received operative orthopaedic treatment and ten have received supporting corsets. All too often an entirely fatalistic attitude is adopted towards patients who suffer from mental deficiency and physical deformities. It is true that the results of treatment are not so dramatic as in those who are mentally normal. It was found however that there were those who wished to be more active, and every effort was made to assist them. Those who could lie, could be made to sit, and were thus given a more interesting world to see. Those who wished to move could be encouraged by physiotherapy, and contracture or muscle imbalance could be diminished. Generally speaking the limiting factor was the mental state of the patient.

The role of operative orthopaedics is, however, limited, and operations must only be undertaken after careful assessment. It must be backed by adequate physiotherapy which is used in encouraging and assisting the patients to walk.

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