

33. The Orthopedic Treatment of Patients with Apallic Syndrome

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Most patients with an apallic syndrome show a characteristic disorder of motor functions which is manifested in an increase of muscle tone and a disorder of body posture, corresponding to the midbrain posture. The upper extremities are thus adducted in the shoulder and flexed in the elbow, while the hand is in a fist position. The lower extremities show a slight flexion of the hip and knee joint, as well as plantar flexion of the feet. But sometimes variations are found that are caused by a lateralization of the midbrain lesion. This again causes an asymmetric posture of the joints, whereby the hip joints are not only flexed but also adducted in one joint and abducted in the other.

These changes of posture are maintained by a centrally regulated increase of muscle tonus. Without treatment a prolongation of these postural anomalies will cause contracture of muscles and a "freezing" of the joints in this position. In the first phase, passive joint movements are still possible; but after shrinking and shortening of the muscles and ligaments, myogenic and subsequent arthrogenic contracture will result. The severity of such an increase of the muscle tone could be observed in one of our patients, who developed a bilateral luxation of the hip as a consequence of the adduction spasm.

A further complication of the apallic syndrome is the development of new bone tissue around the large joints, termed as periarticular ossification. Only a few reports are available concerning periarticular ossifications in connection with the apallic syndrome (Jeannopoulous et al., 1961; Gerstenbrand et al., 1970a), while numerous authors have reported on this problem in association with traumatic spinal cord lesions (Abrahamson, 1948; Armstrong-Ressy et al., 1959; Damansky, 1961; Déjérine and Ceillier, 1918; Liberson, 1953; McNeur, 1954; Miller and O'Neill, 1949; Hardy and Dickson, 1963). But periarticular ossifications were also observed in several other neurologic diseases, such as myelitis (Damansky, 1961), central hemiplegia (Irving and Le Brun, 1954), brain tumor, and meningitis (Lorber, 1953), as well as poliomyelitis (Costello and Brown, 1951; Freiberg, 1952). In the latter, ossifications were also described in the metacarpophalangeal joints. Ossifications of this kind are rarely seen following extensive burns (Johnson, 1957; Evans and Smith, 1959).

Orthopedic Therapy

Therapeutic measures in treatment of the apallic syndrome are initially concentrated on the immediate preservation of life. Even at the earliest stage the orthopedist has to be consulted. The prevention of decubitus ulceration and joint contracture is of greatest importance during the intensive therapy period, and depends on a disciplined routine of nursing management,

which includes turning the patient at regular intervals of 4 h. During the initial stage, i.e., the phase of the acute midbrain syndrome, the patient reacts to the slightest external stimuli with stretch cramps and an emergency reaction which stresses his circulatory and metabolic system. Early attempts at passive joint movements are rejected by the patient's vegetative lability. Passive joint movement should be started and later intensified, only after subsidence of the acute disinhibition of the muscle tone. Before that, special physical therapy has to be commenced by using the tonus-regulating reflexes. In the subsequent course of the remission stage, physical treatment gains in importance.

If, in spite of these measures, joint contractures develop, different orthopedic measures must be undertaken. In addition, it is of the greatest significance to consider the patient's general condition as well as his trophic situation. Premature active orthopedic treatment using casts or bandages, may lead to formation of decubital ulcers on the extremities. They will cause unfavorable healing tendencies and retard the course of remission.

Treatment with Casts

The utilization of removable casts enables one to keep an extremity in a certain passive position. Early in the course of the apallic syndrome this aid may be used as a prophylaxis in joints prone to contracture in order to prevent a malposition. In addition, already existing contractures may still be treated in this manner. A succession of casts are used. The procedure is as follows: A joint inclined to contract is passively extended after which a temporary cast is applied. After an interval of several days this procedure is repeated until correction is achieved. Finally, during mobilization of a patient, these casts may be used for keeping joints within the allowable maximum flexion and extension. Applications of this kind require little effort and since they can be removed, the casts will not interfere with the patient's nursing care. Even physical therapy may be continued without interruption. Pressure points must be watched carefully since decubital ulcers may develop. Cast treatment will be unnecessary after the patient has regained active muscle function.

Wedging

Existing myogenic contractures or already developed arthrogenic contractures on a single joint, may be treated with a wedged cast. Initially a circular cast is applied around the joint to be treated. Transsection of the cast and the maximum possible extension of the joint follow. Then the cast is fixed in this position. Within a few days the contracted tissue will adapt to the temporary extension, after which the above procedure may be repeated.

Redressment

In some rare cases of an apallic syndrome, redressment may offer a further possibility of mobilizing a joint fixed in a malposition through a forced joint movement under narcosis.

Gentle handling is most important to avoid fractures of the usually porous bones. The corrected joint is again kept in casts of plaster.

Tenotomy

Tenotomy may be undertaken if it is impossible to correct a myogenic contracture with one of the above-mentioned methods. Here the tendinous part of the shortened muscle is transected in a Z-shape. After the joint position is corrected, the incised portions of the tendons is adapted and the extremities fixed in a cast. This procedure is especially suited at the knee joint and the Achilles tendon.

Periarticular Ossification

Periarticular ossification may present a therapeutically difficult problem, and this complication is therefore given special attention. In more than 10% of patients with an apallic syndrome, periarticular ossification may develop. Etiologically diverse factors have been made responsible for this; however, none have given a satisfactory explanation.

The most frequent site of ossifications is the elbow, followed by the hip and knee joint. The shoulder is hardly ever affected, and we have never observed changes of other joints.

Clinical manifestations of ossifications are first apparent through limitation of movement of the affected joints. Palpation of the region of the joint reveals firm to bony resistances, clearly demarcated against the surrounding tissue. This is connected with limited ability of movement and atrophy of the muscle. Besides the new bone formation, muscle contracture will mean additional enhancement of the motion disability.

The extent of new bone formation may vary greatly, from small osteophytes that cause only a moderate degree of limitation of movement, to bone bridges which encase a joint and cause complete stiffness. Real osseous ankylosis occurs only rarely.

Changes are radiographically seldom apparent before the 4th week of onset of the disease (Hardy and Dickson, 1963). First, vague and cloudy inhomogeneous shadows are found which gradually gain in density. Further in the course of the disease, the initially diffuse outline will become distinct from the surrounding tissue until finally the bone bridges are radiographically recognizable as highly differentiated bone. Cessation of new bone formation is best seen in serial radiographs. If no further enlargement of the bone bridges with regard to their extension is observed, the end of the ossifying process may be assumed. This process usually takes 7-8 months (Miller and O'Neill, 1949; Hardy and Dickson, 1963).

Morphology

Principally, all tissue originating from the mesenchyma may be drawn into the ossifying process. The transformation takes place first in the periarticular tissue and later in the

neighboring muscles. The joint capsule stays unaltered for some time and shows only degenerative changes. In persistent periarticular ankylosis of the joint, all structures of the joint, including the cartilage, are drawn into the ossifying process. The newly formed tissue finally shows all characteristics of a highly differentiated bone (osteons, blood-building tissue, yellow marrow) (Déjérine et al., 1919).

Indications for Operation

The timing of operation of periarticular ossifications is important. Removal of bone too early may result in a recurrence of ossification even though the operation seems at first satisfactory. The functional ability of the muscular system controlling the joint is of significance. Meticulous topographic knowledge of the ossifications and their relation to vessels and nerves is a prerequisite of any operation of this kind. Finally, the active restoral of function of the extremity being treated must be reasonably related to the extent of the operation to be performed. The goal of treatment of patients with apallic syndrome as compared to patients suffering from spinal cord lesions should not only be in the improvement of the position but also in the regaining of active mobility of a joint.

Operative Therapy of Periarticular Ossification

A radical excision of the bone bridges, independent of the affected joints and the extent of the ossification, is the goal of treatment. This is especially important at the cartilage-bone border. Intraoperative, after the new bone is chiseled off, joint movement must be extensively free. Preparation of the elbow joint may cause trouble because of the sheathed-in arteria cubitalis or nervus ulnaris. In the knee joint great care must be taken to avoid damage to the remnants of the collateral ligaments because their debilitation may cause joint instability. The removal of bone bridges surrounding the hip joint may bring difficulties, for this joint may be completely enveloped by ossified tissue.

Results of Operated Periarticular Ossifications

Surgical experiences of apallic syndrome patients with periarticular ossifications are absolutely encouraging when the operative indication was strictly performed and the operation properly timed. Best results and greatest increase of mobility were attained after removal of bone bridges of the elbow joint (authors' experiences from 6 operated joints) (Fig. 1a, b). The range of motion attained primarily after operation was maintained for 6 years. Operative intervention, other than exact joint rectification, such as plastic operation of the elbow joint, are of no value and must be looked upon as overtreatment. Operative interventions on hip joints with great hindrance of mobility may achieve significant results, even though they are technically difficult (three operated hips). Especially in bi-



Fig. 1a

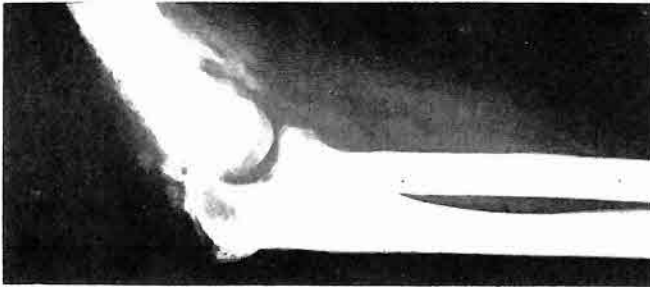


Fig. 1b

Fig. 1. Traumatic apallic syndrome, periarticular ossifications, left elbow with marked formation of bone bridges, dorsal. Patient P.C. male, 40 years old. (a) Before operation, (b) 5 months after operation. Corresponding to operative report joint capsule was mostly unaltered, ulnar nerve sheathed by bone tissue



Fig. 2a

Fig. 2. Apallic syndrome as sequela of massive hemorrhage of corpus callosum; periarticular ossification of both hip joints with total ankylosis. Patient E.M., female, 24 years old. (a) Before operation, (b) 2 months after resection of femur head and collum, left side, with satisfactory function, moderate recidivism



Fig. 2b. (Legend see p. 212)

lateral ankylosis (Fig. 2a, b), surgical treatment will facilitate the patient's ability to walk and sit. Regarding postoperative results, the knee joint is the least amenable; the only indication for operating is bilateral involvement (three operated joints). All operative interventions will only then give satisfactory results if intensive and long-term physical exercise can be guaranteed. Physical exercise should be commenced a few days after operation and possibly be carried on for several months.

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