

ARTHROGRYPOSIS - A PHYSIOTHERAPY OVERVIEW
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The physical characteristics of arthrogyrosis are:-

- Deformed rigid joints
- Atrophy/absence of muscle groups
- Dislocations
- Intact skin sensation

All children are different and need a full assessment and individual treatment.

Typical patterns of deformity:-

- Hands may be clubbed or clawed (ulnar deviation)
- Elbows may be in fixed flexion or extension
- Shoulders - adducted - internally rotated, with 'tipped' hand
- Hips may be adducted, abducted or dislocated
- Knees - flexed or extended
- Feet - talipes
- Spine - Scoliosis

The prognosis for the child with arthrogyrosis is generally not apparent at birth. If there are good dermal ridge patterns then this indicates that there will be good hand function in the future. The achievement of independent walking depends mainly on the on-going development of the nervous system - the extent to which interconnections between cells develop.

The 'underlying' type of arthrogyrosis may be mainly due to deficient functioning of the nervous system - i.e. Neuropathic Arthrogyrosis, or the pathology of the muscle fibres - i.e. Myopathic arthrogyrosis.

The motivation of the child itself plays a significant part in the outcome.

TREATMENT

Physiotherapy should start as soon as possible after birth and continue several times a day during the first year or until they are standing. The aims of the physiotherapist are to correct and maintain the position of the limbs and to preserve the alignment of the joints. It is also important to promote the normal developmental progress to try to achieve skills such as rolling, sitting up and standing at an age appropriate time. The final aim is to achieve independent, low energy walking where possible and, in the case of upper limbs, independence skill such as feeding and self-care.

MODALITY OF TREATMENT

The physiotherapist will begin with passive stretching of tightened soft tissues and gentle mobilisation of joints. They will encourage active limb and trunk movement and generally assist the child to reach the normal milestones. Physiotherapy stretches joint and skin in readiness for operations. Progressive splinting will also be used to maintain position and range of movement of a joint and also to promote independence. The physiotherapist works towards building up a stable trunk which is the key to independent walking. Aids such as rollators and elbow crutches will be introduced when appropriate. Prone lying helps to maintain good hip extension.

Strapping rather than plastering is the preferred option as it allows mobility and normal sensation. Serial splinting should be undertaken with care so as not to damage the nerves and blood vessels.

It is important for the child to learn to tolerate the splints before applying pressure, so we take things slowly to begin with. Splintage and stretching are essential after any surgery

Play has an important role in the development and treatment of the children. Toys that enable the child to move around independently, explore and learn about their surroundings should be provided and adapted as necessary.

SPLINTS

The joint should be splinted daily to maintain or gain a gradual increase in the range of movement. It is generally better to use night splints, especially for the upper limbs, to allow the child freedom of movement during the day for them to feel and play. Velcro to fasten splints is preferable to bandaging them on and toilet paper can be used on the inside of the splints to absorb sweat.

For hand splints a material that can be moulded easily, such as the dental materials Blendagon or Permagon are ideal. The wrist should be corrected first and then the fingers. The wrists need to be slightly flexed, rather than extended in arthrogyrosis to allow better movement of the fingers.

Leg splints can be made from a wide range of materials which can be heat moulded; gaiters and inflatable splints can also be used. To help with walking the child may have callipers or use a walking aid such as a rollator or elbow crutches. Some children use a swivel walker or RGO (reciprocating gait orthoses) to help with mobility.

For babies with dislocated hips there are special splints such as the Van Rosen or Parvick which help to maintain a good position. Children who have poor protective responses because of fixed flexion or extension of the arms, may benefit from having the protection of a helmet.

Regression of joint range will always be a problem until the child stops growing. The soft tissues contract, and go on contracting, while the child is growing. Splints should, therefore, be worn for as long as possible to minimise deformity. Splinting, although beneficial does not replace the necessity of daily stretching and exercise.

SURGERY

The aim for lower limb surgery is to obtain stability and symmetry. Surgery of the feet to make them plantigrade i.e. flat to the floor, should be done before the child is standing (pre-18 months) so that the child maintains the corrected position by body weight when they are standing. The aim for the hips is to achieve reciprocal movement and for the knees to abolish excessive flexion contracture.

The aim of surgery on the upper limbs is to obtain a suitable position for function and to facilitate daily living activities such as feeding, dressing and toileting. To be able to use crutches the child must be able to extend the arms and may require surgery to enable them to do this.

WALKING AIDS

It is preferable to encourage the child to manage with no walking aids unless absolutely necessary as they restrict hand use and encourage flexion of the spine and hips. It is better to encourage a child to walk holding someone's hands which can gradually reduce the support than become dependent on a rollator etc.

PHYSICAL ACTIVITIES

There are numerous benefits in a child with arthrogryposis taking part in physical activities. It can maximise and maintain the available muscle strength and improve stability.

Swimming needs trunk strength but limited limb strength and flexibility. It is non-weight bearing but aerobic, the use of flippers can help with swimming or be used as an exercise tool. It is better that the water is warmer than the average pool.

Cycling is possible for a number of children and there are adapted bikes and trikes including hand-propelled machines that a child can use.

For non-walkers in particular, horseriding gives a great feeling of independence - and superiority! Special saddles or a side-saddle may be needed but local branches of Riding for the Disabled will be able to provide classes and information.

Any aerobic exercise will be good for a child who is not very mobile and in some cases helps with weight loss. Finally, and as important, are the emotional and social benefits. The increase in self-esteem and the mixing and making of friends in a group, plays an important part in the overall development of the child.