Physiotherapy for Children with Arthrogryposis

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The management of musculoskeletal problems associated with Arthrogryposis Multiplex Congenita (AMC) can be challenging; therefore, we feel these children are best managed by multidisciplinary teams. The child, parents and family are the most important members of that team and without their dedication and support; physiotherapy treatment would not be successful.

Babies

When a baby is born it should be assessed as soon as possible after birth. A respiratory assessment is occasionally needed as chest physiotherapy may be indicated to maintain a clear chest. Passive range of movement is recorded at all joints and muscle strength is measured using the Oxford scale which grades muscle power between 0-5. In babies, a reliable record is virtually impossible so muscles are recorded if they appear to be working and if they can work against gravity. Babies are observed in a variety of positions & by palpating muscle contractions. Physiotherapy is more intensive in the first year, and then decreases during school years.

Stretches

A programme of passive stretching is carried out for all joints with contractures even if there is no movement in a joint. In order to be effective, they may be a little uncomfortable but should not be painful. The first three months are particularly effective but consistent stretching is most critical during growing years and especially within the first two years of life.

The physiotherapist should teach the parents so they can continue the daily programme. The frequency of stretching should be 3-5 sessions a day initially with 3-5 repetitions of each stretch, holding each stretch for 20-30 seconds. It is encouraged that stretches are incorporated into a routine, for example, lower limb stretches can be carried out with nappy changes and upper limbs when clothing is put on or removed.

Positioning

Positioning is an important tool to stretch joints. It also encourages development of the head and trunk, strengthens limbs & facilitates function. Babies need this head and trunk control to progress through their developmental milestones. Prone lying helps stretch hip flexion contractures and side lying helps to bring hands together. Corner chairs can provide support for a child who is unable to sit independently, allowing them increased stability to play.

Splinting

Splints maintain stretch with the limb in a comfortable position. Recommendations for duration of the time spent in the splint vary from at least 6 hours out of 24 hours to maintain range, to 22 hours a day. Tardieu (1988) reported no progressive contracture when the soleus muscle was stretched for 6 hours a day in Cerebral Palsy over a 7 month period, whereas Donohoe and Bleakley (1994) recommended 22 hours a day in AMC to be maximally effective. I would recommend splinting more frequently in the first 3 months whilst it is not interfering with function, allowing some time without splintage for the baby to develop muscle power. After the first 3 months, day splints should only be used if a child's function is improved in the splint, and night splints should be used for stretching. In the upper limbs, elbow flexion splints are worn during the day if they allow for a more functional position for play activities, and extension splints are worn when the child is asleep. Serial splints help to open up hands at birth and again functional day wrist splints should only be worn if they improve the child's hand function. Night wrist splints usually need to include the

fingers in order to stretch them. In the lower limbs, splints in this age group are standing splints and ankle-foot-orthosis (AFO'S).

Casting

A common foot problem in AMC is the clubfoot deformity. The most successful treatment for this deformity in the idiopathic newly diagnosed clubfoot is the Ponseti technique. It was devised by Professor Ponseti in the 1940's & he is still practising in Iowa at the age of 93 years. The trend in Britain towards surgical management has reversed during the past decade for idiopathic clubfeet, with the Ponseti method of serial casting followed by bracing now being the first line management. Most idiopathic feet (95%) treated by this method achieve an excellent result. It is inexpensive, effective and feet studied in a 35 years follow-up study by Professor Ponseti are strong, flexible and pain free. Idiopathic clubfeet are normally corrected within approximately 6 weeks, by weekly manipulations followed by long leg plaster casts. A percutaneous tendo-achilles tenotomy is often needed and this is followed by a cast for 3 weeks. Once the corrected foot position is achieved, a foot abduction brace is worn full time for 3 months and then 14 hours a day until the child is 3-4 years old, to maintain the position.

Arthrogrypotic clubfeet are known to be the most resistant to treatment. Although the use of Ponseti in the AMC clubfoot has not been reported in the literature to date we have had some success with this technique in AMC. Two to three percent of idiopathic clubfeet are difficult to treat & described as atypical/complex clubfeet. I surmise a larger percentage of Arthrogrypotic clubfeet fall into this category and this requires a modification of the standard Ponseti treatment in order to succeed. The Ponseti technique has had a huge impact on the treatment of clubfoot and in my opinion all AMC babies with a clubfoot should be referred for Ponseti, but this referral should be to a specialist centre with experience of atypical feet and the modified technique.

Toddlers

Moving on to toddlers, children need to be re-assessed continually. Ability rather than disability should be stressed with a strong emphasis on assisting the child through problem solving, enabling independence. Passive & active ranges of movement continue to need close monitoring; formal manual muscle testing is important and the emphasis should be on assessing functional muscle strength. AMC children are frequently strong in the mid range but unable to move in the shortened end range. Children should also have a comprehensive gait assessment. This should include distance, speed, step length, gait deviations, use of assistive devices and muscle activity. It is important to determine the level of bracing required and level of independence in self care skills. In this age group, gains in range have usually been achieved and positions need to be held with splintage and positioning to provide adequate stretch. Often night splints for legs and hand splints are needed.

Treatment for toddlers needs to continue but less frequency may be more realistic. Disruption to the child's and family's life needs to be minimized: time in therapy is time not spent on other activities i.e. education, socialization and play which are also really important for the development of a toddler. Stretches should be carried out once to twice a day. These children should now be mobilizing independently, with or without splints and aids and if this is not possible, standing should be initiated in a standing frame. By the age of one, a child should tolerate one to two hours a day in a standing frame as this helps self-stretching of the feet and leg muscles. Mobility should be maximized with the minimum bracing and use of assisted devices. Walking aids may need to be customized to accommodate upper limb limitations. Splintage should be lightweight and can be articulated or fixed. It is preferable if the child can put on or remove their own orthoses.

The child's function in activities of daily living challenges should be improved. Poor upper

limb function from the contractures and lack of muscle power limit the child's independence in feeding, dressing etc. This can be a shock to many parents when the child is no longer an infant in whom dependency is expected. Difficulties with upper limb function are often overcome with trick movements. The aim is for independent feeding and toileting, and if personalized equipment is needed a referral to occupational therapy may be necessary. Physiotherapy should concentrate on increasing strength by progressing functional activities and this can be addressed through play.

School Age Children

Some children, as they grow older and enter school age may need an electric wheelchair to keep up with their peers. Cumbersome bracing, inefficient gait and poor upper limb function can limit a child's ability to participate in the playground or social activities. When one considers speed, safety and energy conservation it may be better to supplement mobility with a wheelchair.

Children should be responsible for their self-care and exercise programmes. Physiotherapy treatment focuses on assessing children in the classroom to ensure the school environment is adapted as much as is feasible to suit the child - for example, desks are the correct height and angle, as minor alterations can make a huge difference to independence.

As children become bigger and heavier with age they may find walking more difficult and they may lose movement at joints, despite stretching throughout the growing years. Surgical correction may be necessary to improve range. Bursts of physiotherapy may be required after surgery or targeted stretching programmes needed for specific joints.

A study looking at knees and function in teenage children with AMC concluded that children who were born with flexed knees often preferred to be wheelchair bound in their late teens; approximately fifty per cent of these children were walking independently in the community. In contrast, those children born with extended knees had a better response to physiotherapy and stretches, and the vast majority were able to walk independently in the community (Murray & Fixsen 1997).

If a child requires surgery, he/she may need a physiotherapy assessment to help establish muscle power, function, range of movement and whether that range is in the most functional position. Overall function needs to be assessed i.e. what a child will gain or lose by undergoing surgery. Surgeons may be able to give a child the ability to eat independently but they may be taking away the ability to stand from sitting independently if a child needs two arms extended to achieve this. Overall function must be improved; these functional assessments are carried out when the child is old enough to assist in these difficult decisions.

Children with Arthrogryposis are motivated and rewarding to know and to treat. I think physiotherapy has a vital role to play in their lives and in my experience early assessment, treatment and management is essential to improve their quality of life.