

Cerebral Palsy

Dystonia in children with cerebral palsy

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Cerebral palsy (CP) is the most common cause of physical disability affecting children in developed countries occurring in about two per thousand live births. CP is an umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of its development.

Cerebral palsy can be described in terms of the type of movement disorder children present with. The most common movement disorder is spasticity (present in up to 88% of people with CP). Dystonia is another type of movement disorder which occurs in up to 15% of children with CP. Some children have both spasticity and dystonia. Dystonia is characterised by involuntary muscle contractions that may affect one or more limbs, or be present throughout the child's whole body including their mouth, eyes and neck. Involuntary muscle contractions cause twisting or repetitive movements and abnormal posturing of the affected body part(s), and are often painful and very distressing for the child.

Practical consequences of dystonia for the child include: 1) difficulties in completing self-care tasks, for example, dressing and eating, 2) problems accessing a computer for schooling or leisure, 3) difficulty with mobilising safely and 4) difficulty being comfortably seated in a wheelchair. These difficulties can lead to limitations in participation and reduced quality of life for the child, which in turn have negative consequences for the parent(s)/caregiver(s).

Medication is frequently used for dystonia, but little information is available as to whether it is effective, and side effects are common. Most of the published studies assessing medications are small and descriptive, limiting the conclusions that can be drawn from them. One of the most commonly used medications is benzhexol hydrochloride, also known as Artane (trade name) or trihexyphenidyl. Benzhexol hydrochloride is an anticholinergic agent with a long history of use in the management of extrapyramidal disorders in children and adults. It has a relaxing effect on smooth musculature, exerted both directly upon the muscle tissue itself and indirectly through an inhibitory effect upon the parasympathetic nervous system.

One study reported on a prospective, open-label, multicentre pilot trial of high-dose benzhexol hydrochloride in 23 children aged 4 to 15 years. The children had CP with secondary dystonia which was impairing function in the dominant arm. There was variability in children's response to the medication. On average the group showed improvements in arm function after 15 weeks of receiving benzhexol hydrochloride compared with baseline, however further analyses showed that a subgroup of children with hyperkinetic dystonia worsened over this period (Sanger et al 2007). The authors concluded that the evidence for using this drug to reduced dystonia in CP remains equivocal and that larger trials are required. A small randomised double-blinded, placebo-controlled, cross-over trial of benzhexol hydrochloride with 16 children found little evidence of a difference in the primary outcome measure, the Barry-Albright Dystonia Scale for global assessment of dystonia (BAD

Scale). Secondary outcome measures included the Quality of Upper Extremity Skills Test, the Canadian Occupational Performance Measure (COPM) and the Goal Attainment Scale (GAS) (Rice and Waugh 2009). Although there was no treatment effect for the group overall, a greater proportion of children in the treatment group showed a clinically significant improvement on these outcomes in the active phase of treatment compared with the control phase. Importantly though, side effects were common while receiving benzhexol hydrochloride.

In 2011, Carranza-del Rio et al undertook a retrospective chart review of 101 children treated with benzhexol hydrochloride. Ninety-six percent of children demonstrated benefits post treatment compared to pre-treatment including a reduction in dystonia in the arms (59%) and legs (38%), although side effects were reported in 69% of children including constipation, urinary retention, sleep disturbances, behaviour changes and excessive dry mouth.

Despite a small number of studies reporting on the use of benzhexol hydrochloride, there remains some uncertainty about the benefits of using this drug for treating dystonia in CP and further investigations are required.

Rice J, Waugh M. Pilot study on trihexyphenidyl in the treatment of dystonia in children with cerebral palsy. *J Child Neurol* 2009;**24**:176-182.

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Sanger TD, Bastian A, Brunstrom J, et al. and the Child Motor Study Group. Prospective open-label trial of trihexyphenidyl in children with secondary dystonia due to cerebral palsy. *J Child Neurol*. 2007;**22**:530-7.