

Parents with children who have Usher syndrome may find their child presents with difficulties involving their balance, from an early age. The level of difficulty experienced by the child will vary from child to child and may depend on the type and severity of their condition.

***About the human balance system (content below taken from [www.sense.org.uk](http://www.sense.org.uk))***

*This system is the coordination of information from three sources in your body. These sources are:*

- 1. Your visual system*
- 2. Your vestibular system (this is information from the balance and orientation components in your inner ear)*
- 3. Your proprioceptive system (this is information about the position of your feet, limbs and joints)*

### ***Balance and Usher***

*An individual with Usher syndrome will have balance that can be affected by issues with two sources of the balance system - the vestibular and vision systems.*

### ***The vestibular system***

*The vestibular system is within the ear and consists of a series of fluid-filled circular canals set at different angles. When the head moves, the rolling fluid tells the brain exactly how far, how fast and in what direction the head is moving, by relaying the information down the vestibular nerve. If the brain knows the position of the head, it can work out the position of the rest of the body. When the vestibular system does not work the brain actually works hard to compensate for the loss in a number of ways, including making greater use of other senses.*

### ***Usher type 1***

*People with Usher type 1 have issues with balance, primarily because the vestibular system does not work (vestibular dysfunction) from birth. Consequently, babies with Usher type 1 may demonstrate delays in sitting and walking. The visual impairment associated with Usher type 1 then further impedes good balance, especially at night or in poor lighting conditions.*

### *Usher type 2*

*Usher type 2 is not associated with vestibular dysfunction, meaning that balance should be similar to that of people without Usher. However, the visual impairment means there may be some lesser issues with balance, especially at night or in poor lighting conditions.*

### *Usher type 3*

*Some people with Usher type 3 experience balance issues as a result of vestibular dysfunction. It is currently thought that any dysfunction occurs later in life. As with Usher type 2, people with Usher type 3 may experience some problems with balance as a result of their visual impairment, especially at night or in poor lighting conditions.*

*The combination of tunnel vision and night blindness with vestibular dysfunction may make individuals with Usher syndrome more prone to accidental injury.*

It is important that your child is assessed on a one to one basis by suitably qualified and experienced physiotherapist who will carry out a comprehensive and individualised assessment of your child to establish your child's particular functional problems. They will assess the child's individual balance capabilities, (how well they can stand still, walk and balance during functional tasks such as standing on one leg, walking on a narrower edge, reaching forward run jump etc.), assess for any deviation in normal eye movements (controlled by the balance organ in the ear) and assess if the child is meeting their developmental mile stones (can they walk, stand on one leg climb stairs etc. at the correct age). It is the opinion of this author that this person should be a chartered physiotherapist with specialised training in children's physiotherapy and vestibular disorders. The chartered physiotherapist will also assess the other systems that might affect the child's ability to balance such as muscle strength, coordination and range of movement of the limbs – all of which will help the child carry out functional balance tasks.

Assessment of our peripheral vestibular system can be done by an audiologist or clinical scientist in Audiology. This can involve a combination of tests designed to establish how well the inner ear is responding. However the balance system is complex and assessing one system in isolation may not reveal the whole clinical picture.

Once the vestibular dysfunction is diagnosed it is thought best to start rehabilitation as early as possible to optimise balance function whilst vision is still present.

However, it can be very difficult to find such a practitioner within your locality. You can request a referral from your GP or paediatrician, and ask to see a physiotherapist experienced in dealing with vestibular disorders in children. It may help to look up ACPIVR (Association of Chartered Physiotherapists Interested in Vestibular Rehabilitation) or APCH (Association of Paediatric Chartered Physiotherapists) for help in finding a suitably experienced physiotherapist.

Following the initial assessment your child may need vestibular rehabilitation (or balance rehabilitation). Vestibular rehabilitation is a type of exercise based therapy that can help improve functional balance, reduce dizziness and improve gaze stability. Although there is a lot of research demonstrating that vestibular therapy can improve balance and reduce dizziness in adults and children (see the work of RoseMarie Rine, an American paediatric vestibular physiotherapist and researcher), there is no evidence yet that this is effective in children with Usher syndrome specifically.

Once a thorough assessment of your child has been completed, the physiotherapist will liaise with you and your child to compile a management plan based on your particular short-and long-term goals. Treatment may consist of one to one supervised exercises with the physiotherapist but you should be instructed to continue these exercises at home on a daily basis. Activities to challenge and promote balance function will be encouraged and these should be incorporated into the daily routine and play activities.

**Parents should only encourage such exercises once it has been deemed safe to so by the physiotherapist. Exercises should challenge the child's balance system but should be performed in a safe environment with appropriate supervision to limit the risk of falls and injury.**

Children should be followed up regularly to check they are meeting their goals and have their treatment plan progressed or modified accordingly.

**Examples of Exercises (only to be carried out once safe to do so by a qualified physiotherapist)**

Exercise that may help improve balance: games that encourage: standing still like a statue, standing on one leg, narrow walking practice, obstacle courses (stepping over and around objects) walking with quoits on head, egg and spoon walking.

Research shows that in order for each patient to get the best outcomes they should follow an individualised customised exercise program, and so for this reason specific exercises have not been listed here. Instead, please seek the guidance from a chartered physiotherapist with the correct specialism, as referred to above.

**Information sheet compiled by Senior Physiotherapist, Manchester Royal Infirmary, UK**