

# Thalidomide Embryopathy

## Overview of the health condition or disability

*What is the condition usually called?*

Thalidomide Embryopathy

Sometimes known as thalidomide damage.

There are a number of terms that are used when referring to someone who was damaged in utero by maternal ingestion of the drug thalidomide, these include:

- Thalidomide survivor
- Thalidomider

Most people do not like the term 'thalidomide victim' but it would be best to ask the person you are assessing which term they prefer and then use this throughout the assessment.

We also differentiate between upper limb damage and lower limb damage – the most common types of thalidomide damage - and the terms “short arms”, “short legs” are acceptable.

Thalidomide damage is seen as a rare disease, there are very few experts on its effects in the UK other than The Thalidomide Trust who are highly knowledgeable and recognised as the definitive authority.

## Commonly reported variability in functional restrictions

*Do people with this particular health condition or disability find its impact on daily life can vary from time to time?*

Yes

*What aspects of daily living can be worse and what might be constant?*

Some aspects of the impact of thalidomide embryopathy are constant – for example absent or shortened limbs, hands and/or feet, missing digits, hearing impairment, sight impairments and facial paralysis have a continuous impact.

Someone with absent or short arms will constantly have to use their body in a way that it was not designed for – reaching or bending constantly throughout the day and frequently using other parts of their body (such as teeth and feet) to perform tasks generally done with hands. This has a constant impact on their joints and muscles, which are often themselves abnormal as a result of the thalidomide damage. They will always have to develop their own ways of undertaking the activities of daily living and although these have become usual to them, they cause pain and fatigue (the levels of which will vary) and have a significant impact on health and wellbeing over time.

Someone with absent or short legs will have constant mobility issues and may use their arms to move around. Their levels of pain and fatigue from this will vary. They may use a prosthesis and will have similar issues to amputees but these will be exacerbated by the fact that there is no surgically created stump so problems with fit, comfort and use are common and variable. Pressure sores may result from sitting in a wheelchair and/or from prosthetic use.

Someone with facial damage will suffer social anxiety and distress as a result of their highly visible disfigurement.

The psychological distress of being stared at and having photographs taken on mobile phones is constant especially as the thalidomide scandal was such a significant public event. This can limit the ability to travel and socialise.

Severe and chronic **pain** is a significant issue as a result of living with a damaged body. While some level of pain is generally constant, when experiencing pain flare-ups someone with Thalidomide Embryopathy will be significantly less likely to be able to carry out daily living activities. Pain may develop after activity and many Thalidomide survivors can only do a task once, or have to follow any activity with a corresponding period of rest. For example, 20 min of housework with a 1 hour rest. A day of travel with a day or two of rest. **Fatigue** is related to pain and overuse and is commonly experienced.

**Temperature control** is an issue for many thalidomide survivors – they often experience either hyperhidrosis (excessive sweating) or feel the cold excessively. This impacts their ability to function when the weather varies.

**Balance and falls** as a result of pain and fatigue, ear problems (many thalidomide survivors were born with very narrow ear canals) use of prosthetics and/or medication such as strong painkillers. Due to the inability of a thalidomide survivor with absent or short arms being able to save themselves from serious injury when falling or the fear of falling and anxiety is a constant factor.

**Prosthetics** – some thalidomide survivors with leg damage use prosthetic legs. As they have not had a “clean” amputation they are very likely to experience poorly fitting sockets, causing discomfort, pain and insecurity when moving about. Many use them for appearances sake rather than for effective mobility.

**Ageing and the impact of thalidomide damage.** Evidence shows that thalidomide survivors’ health has deteriorated gradually over time. Now they are all in their later 50s/early 60s, they are experiencing a number of age-related health issues (including incontinence, joint problems and problems with balance) and these are exacerbated by the thalidomide damage resulting in a disproportionate impact on their health and levels of functionality.

## Presenting symptoms

Can include, but not limited to any combination of the following:

- **Upper Limb damage** – No arms, arms short above the elbow, arms short below the elbow, hand and wrist damage, missing digits
- **Lower Limb damage** – No legs, legs short above the knee, legs short below the knee, hip damage, foot damage
- **Missing joints** – Knees, hips, elbows, wrists – in shortened limbs
- **Reduced range of movement** – It may not be possible to bend or raise limbs
- **Spinal damage**
- **Absent external ears** – May use prosthetic ears
- **Damaged internal ears**
- **Facial palsy and damaged facial bones** – Small jaw and overcrowding of teeth

- **Vision impairment and sight loss**
- **Hearing impairment**
- **Internal damage** – Bowel, reproductive organs, vascular system, kidneys etc.
- **Stomas** – Resulting from anal stenosis and internal damage
- **Infertility**
- **Pain**
- **Neurological issues** – Numbness, tingling, pins & needles, loss of grip and strength
- **Psychological issues** – Most commonly depression/low mood and anxiety
- **Secondary damage** – **Arthritis**

## Common areas of daily life where functional restriction occurs

- **Cooking and Eating:** Reduced ability to cut, chop, lift, and stir safely. Likelihood of burns due to reduced reach and proximity of hot items to body. May need to have food cut up or be fed. May be unwilling to eat in public or require assistance to do so if would normally use feet to eat at home. Risk of falls if using legs/feet for cooking or eating. Weight management can be an issue due to reduced mobility, pain and low mood.
- **Washing and dressing:** Thalidomide survivors may be unable to reach all parts of their body for washing and dressing due to reduced reach, inflexibility, limited range of movement, poor balance and/or pain. They often develop their own methods which may be unsafe and or cause pain and fatigue – for example putting a sponge on the shower wall and rubbing against it. There is a risk of slipping and falling in shower/bath and a high level of fear of this, so even if can bathe alone may need another person to be in the house. Clothes need to be specially adapted – e.g. shortened sleeves, and easy to wear. Some avoid underwear and bras as they are too difficult to manipulate.

No belts, buttons, zips without help. Slip on shoes or shoes that can be removed easily if feet are used to undertake tasks. Leg damage, will have difficulty transferring and may need to sit on floor in shower.

- **Toileting:** Transferring, cleaning themselves, need extra space for a prosthesis or wheelchair, may have an adapted toilet at home. They may also have a lot of difficulty cleaning themselves after an 'accident'.
- **Communicating:** Some thalidomide survivors have a hearing impairment as a direct result of their thalidomide damage. It is also a very common feature of thalidomide damage to have narrow ear canals which frequently become blocked with wax, affecting hearing. Inserting and removing a hearing aid is very difficult for someone with short/no arms. Using sign language is also difficult for someone with arm or hand damage. Sight impairment may result from facial damage and putting on and removing glasses/contact lenses is very difficult for those with short/no arms. Facial disfigurement may also cause speech impairment. There are high levels of social anxiety and distress as a result of being stared at. For those with short/no arms, there is a limited ability to gesticulate with hands. Many thalidomide survivors had interrupted education as a result of repeated surgical interventions and there is some evidence of learning disability and autism amongst the thalidomide group – this has often gone undiagnosed and unsupported because the focus has been on the physical disability and the expectations have been low for some survivors.
- **Reading:** As all thalidomide survivors were born between 1959 and 1965 they often had a poor educational experience as a result of frequent hospital appointments disrupting their schooling and low expectations of children with such severe disabilities. Holding a book and turning pages is difficult with short arms. It is often not possible to hold a newspaper. Wearing glasses for reading is not always possible for those thalidomide survivors with facial damage and damaged external ears.
- **Engaging with others:** There is increasing evidence of mild to severe social phobia and horror of being stared at or photographed which has become more pronounced with the introduction of camera phones and social media. As a result will restrict going out to only where they are known, others may not go out at all and some only go out accompanied. High levels of anxiety and feelings of physical vulnerability can limit

thalidomide survivors' willingness to engage with others. For those with speech and hearing impairment there is emotional vulnerability arising from being unable to make themselves understood.

- **Make budgeting decisions:** The high levels of depression and low mood may mean that some people make unwise decisions as a result many thalidomide survivors rely on support from the Trust to manage their money, plan a budget and make financial decisions.
- **Planning and following the route of a journey:** High levels of anxiety can be present, as a result many thalidomide survivors do not go out alone. Routes must be planned to provide access to a toilet they can use safely (for both lower and upper limb damage). Hearing impairments can cause missed announcements and therefore being diverted / getting lost and not being able to ask or understand directions. Sight impairments mean that some thalidomide survivors are completely reliant on help with this aspect of life.
- **Driving:** Thalidomide survivors are often very reliant on a car as their sole means of maintaining independent mobility, as unable to walk any distance or use public transport safely. May still find driving tiring or painful and not be able to drive long distances due to pain or toileting issues. May have adapted car but some are able to use automatic vehicles with electronic controls – e.g. automatic lights, keyless entry, automatic boots and rain sensors. May need to be accompanied on journeys as they will need help to open doors etc. Due to anxiety, thalidomide survivors may only be able to drive to familiar places. If short/no arms may not be able to use ticket barriers, parking machines etc.
- **Walking** (both indoors and outdoors): Thalidomide survivors with leg damage may be unable to walk outdoors and will rely on wheelchairs. They may move around on the floor at home by crawling or walking on their hands. This causes pain and stress on the shoulders. Thalidomide survivors with short/no arms may be able to physically walk outdoors but this is often limited by pain in the hips and knees caused by overuse of these joints to compensate for upper limb damage. Abnormal gait is common which causes back pain after short periods of walking and balance issues and they may be unable to use usual aids and equipment such as walking sticks due to short arms or poor grip.

Fear of falling – due to an inability to break their fall - means they may not go out alone, or in bad weather and only on familiar routes. Many thalidomide survivors struggle to use stairs due to problems with their balance and fear of falling.

*What areas of daily life may a person with this particular health condition or disability find challenging?*

Thalidomide survivors are used to finding all aspects of daily life challenging and have developed their own ways of managing, for example using their feet instead of hands or vice versa. As people age these adapted ways of managing are becoming harder – they are losing flexibility and will be experiencing increased levels of pain that has become chronic.

Even where this is the case, there is a fierce resistance to change despite the fact that these activities cannot be said to be being undertaken safely or reliably, in a reasonable time or to an acceptable standard. Functionality is deteriorating in all thalidomide survivors and there is no prospect of improvement.

*Are there any areas that a Health Professional should ask about specifically to ensure a complete report?*

- The impact of any internal damage that is not apparent to the assessor
- The use of prosthetics – including ears - how these are managed and any associated problems
- The levels of pain and fatigue that are experienced, how these vary and impact on ability to carry out tasks of daily living on different days
- How long it takes to carry out each activity when it is done in a unique or adapted way – it may take five times longer for a thalidomide survivor to make a sandwich for example
- What assistance is provided and relied on by any family or friends – the extent to which this informal care is relied on is often not recognised
- The impact of low mood and anxiety on motivation and the ability to undertake activities of daily living – especially in respect of mobility. This may be associated with alcohol misuse.

### **PIP specific questions – Where to probe at assessment:**

- Consider the limbs effected and ensure you consider these when asking questions in the functional history. The limbs effected will alter your questioning techniques and if multiple limbs are absent, this all needs to be considered.
- If the claimant has adapted and has techniques to enable them to complete an activity as many people with this condition do, consider STAR. Are they able to do this safely, timely, to an acceptable standard and repeatedly? If not, why not? What is it that means they are unable to complete it in such a way?
- Ensure you probe around the other comorbidities that can be affected by this condition that aren't necessarily so obvious such a hearing and vision loss and impacts on mental health. If the claimant is effected in this way, ensure you sensitively probe to gather a robust functional history and remember to do additional examinations such as hearing, vision and the MSE if appropriate.
- Pain and fatigue associated with the condition needs to be appropriately explored and repeatability alongside the other aspects of STAR considered.
- All the other usual questions in the SOH will also be beneficial in such a case around work, home life, hobbies and interests and driving, so please remember to adequately but sensitively probe.
- Probe sensitively if the claimant has a prosthetic limb. Does the claimant have any difficulties in wearing this? Can they only tolerate the limb for a certain amount of time? If so, why is this and how often does it impact on them wearing the limb?
- If they have a prosthetic to the lower limbs, are they at risk of falls? Have they fallen? How frequently does this occur? Have they required a hospital attendance or admission due to injuries?
- A12 – Ascertain how far the claimant can walk and what places they are able to go to. Do they do this unaided or aided? Does the exercise tolerance change on a daily basis? What is the minimum and maximum they can walk reliably on the majority of days?

### PIP specific – Points to consider:

- Consider the information within this document about the journey a claimant with this condition. If they aren't taking any pain relief, ask why and consider this when making decisions on descriptor choices.
- Remember STAR! As with any case, this is very important.

### Common misconceptions about the health condition or disability

It is really important to be aware of the historical context of thalidomide. There can be a misconception that the damage is caused by a genetic condition or acquired disability and the fact that some clinicians and other professionals appear to be unaware of the Thalidomide scandal does not get any assessment off to a good start.

Thalidomide was a significant scandal in the early 1960's and the children born damaged as a result have been in the public eye for most of their lives. As children they were objects of interest and enquiry to medical professionals and social workers, many were given up by their families on the advice of those professionals and as a result there is a commonly held distrust of scrutiny and assessment. This is likely to lead to thalidomide survivors being anxious or uncooperative during an assessment.

*What kind of areas might a claimant with this health condition or disability find hard to mention or perhaps understate the importance of?*

Thalidomide survivors have been trailblazers for independence and achievement in the face of their disabilities. They were told they would not survive childhood, then would never work and not marry or have children. One by one they have defied these predictions and they are fiercely independent as a result and many are unwilling to admit when they are not coping or are finding day to day tasks difficult.

Recognising that the way they usually carry out activities of daily living may now be having a negative impact on their health and can potentially be unsafe, is really difficult because they may fear the total loss of their independence. They may understate the difficulty or safety of how they have to manage on a day to day basis.

One example is toileting where some were taught at a young age to put the paper on the toilet seat, a cabinet or the floor and rub to clean themselves. They may find this difficult to mention and when challenged will say they are able to do clean themselves properly despite the fact that they have to rely on a shower to clean themselves afterwards. If they are using this method it should be accepted they cannot clean themselves properly.

*What areas of this health condition or disability are least well understood or hardest to identify and which might impact on daily living?*

Having been damaged by a drug, many thalidomide survivors do not like taking medication and may be unwilling to have pain relieving injections or surgery. This means they live with high levels of pain and may be so used to this that they may not mention it appropriately.

Many thalidomide survivors experience difficulty monitoring medical conditions. For those with no arms there may be an inability to measure blood pressure accurately and have blood taken. Monitoring blood sugar can be a problem for those with diabetes. It is not always easy to get medication doses correct for someone with absent limbs. As a result there may be a lot of variation in symptoms from day to day which impacts on activities of daily living. The psychological impact and distress from the risk and fear of falling. Even if a thalidomide survivor has not yet had a serious injury, not being able to put your hands or legs out to prevent a fall or protect yourself is very frightening.

Complex pattern of damage in an upper limb can prevent the limb achieving a certain movement required to undertake activities of daily living. A damaged limb of whatever length is likely to have a limited range of movement in the shoulder, elbow and wrist together with a reduced grip.

The impact of secondary damage that has been caused as a result of using the body in a way for which it was not intended. For example, to compensate for a reduced reach overuse of the neck, shoulder and spine is needed. Using arms to move around because there are no legs present causes severe stress on the shoulders, hands and/or elbows, resulting in pain, arthritis and joint degeneration. However the thalidomide survivor has no alternative and must continue to make these painful movements, despite the medical view that pushing past the pain should be avoided as this damages further the misaligned joints, can cause muscle spasm and can exacerbate the pressure on the normally formed joints.

### *How is it best to ask about these areas?*

Make it clear that you understand the history and context of thalidomide and understand that they are survivors of medical damage. Acknowledge the high level of independence that they have achieved but also acknowledge that you understand the impact of them using their bodies in ways they were not designed for and the impact of ageing – creating a safe space for them to be honest about any things they are finding difficult.

## **Recommended communication approach to a claimant at a face-to-face consultation**

*What are the best communication approaches necessary to engage someone with this health condition or disability?*

- Recognise that some people are very anxious and some will be angry that they have to go through the application process when it has already been acknowledged that they have been damaged through no fault of their own (and are likely to have been previously awarded DLA for life). Be patient and courteous even in the face of this.
- Be open and honest. It's ok to ask about the extent of the limb damage and any other unseen damage.
- Be personable and friendly, but professional in order to provide confidence that you are able to understand the extent of the impact of living with thalidomide damage.
- Ask follow up questions to explore how someone would undertake the activity you have asked them about.
- Be very clear about the point of the question – for example one thalidomide survivor was asked “can you use a knife?” to which he replied “yes”. Then “can you use a fork”, again “yes”. So the assessor recorded “can use a knife and fork” – despite the fact that the person being assessed had only one short arm and was not able to use both utensils at the same time.

## **Etiquette and common courtesies**

*Is there anything that would make the claimant feel more at ease?*

- Look – is there a hand to shake? It's ok to ask if a handshake is welcome – don't assume it is not possible.
- Make it clear that the assessor understands the context of thalidomide and that it is not a disease or an acquired condition.
- If there is water in the room make sure it is within reach of short arms (don't put things in the middle of the table). If possible provide drinking straws.

## **Further reading:**

The Thalidomide Trust Upper Limb Damage Impact statement - <http://www.thalidomidetrust.org/wp-content/uploads/2017/03/Upper-limb-impact-statement.pdf>

The Thalidomide Trust PIP case studies:

- <https://www.thalidomidetrust.org/wp-content/uploads/2019/01/Case-Study-1-Upper-limb-and-eyesight-affected-1.pdf>
- <https://www.thalidomidetrust.org/wp-content/uploads/2019/01/Case-Study-2-Upper-limb-and-back-affected.pdf>
- <https://www.thalidomidetrust.org/wp-content/uploads/2019/01/Case-Study-3-Upper-limb-affected.pdf>
- <https://www.thalidomidetrust.org/wp-content/uploads/2019/01/Case-Study-4-Hearing-impaired.pdf>
- <https://www.thalidomidetrust.org/wp-content/uploads/2019/01/Case-Study-5-Lower-limb-affected.pdf>
- <https://www.thalidomidetrust.org/wp-content/uploads/2019/01/Case-Study-6-Upper-limb-ear-and-eyesight-issues.pdf>