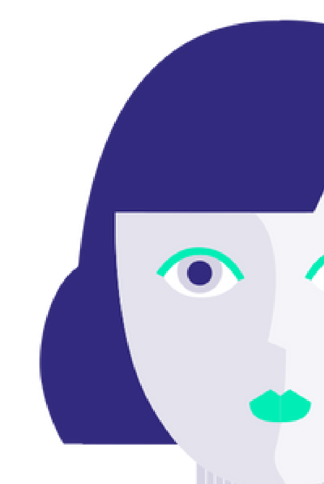
Aspiration in Rett syndrome

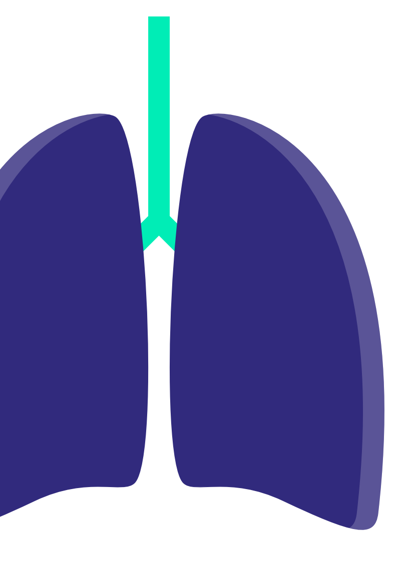
Keeping someone with Rett syndrome safe from aspiration



Lower respiratory tract infection is the most common cause of death in Rett syndrome, followed by aspiration/asphyxiation (31.6%), respiratory failure (14.0%) and seizure related illness (5.3%). [i]

Often, lower respiratory tract infections/chest infections and pneumonia are caused by aspiration.

Aspiration means breathing foreign objects into your airways.



Usually, it's food, drink, saliva, or stomach contents when someone swallows, vomits, or experiences reflux.

Most of the time, for most people, aspiration won't cause any symptoms. We all have had the experience of something we're eating or drinking 'going down the wrong way.' Most people can usually cough out a foreign object before it enters the lungs.

But aspiration is more common in older people, infants, and people who have trouble swallowing and it is dangerous.

Simply put, the hatches which deal with food/drink and breathing are very close to one another and the way our eating and drinking and breathing systems work alongside each other is complicated.

Problems with coordinating swallowing and/or breathing can lead to aspiration which can lead to chest infections/aspiration pneumonia.

Analysis of information from death certificates via the Confidential Inquiry into Premature Death for People with Learning Disabilities **(CIPOLD)** [ii] has shown that people with learning disabilities are much more likely to die of the consequences of solids or liquids in their lungs or windpipe than those in the general population. The analysts concluded that this is a common, *possibly preventable cause of death.*

Here are some signs that a person may be aspirating:

- arching or stiffening of the body during feeding
- irritability or lack of alertness during feeding
- refusing food or liquid
- failure to accept different textures of food (e.g., only pureed foods or crunchy cereals)
- long feeding times (e.g., more than 30 minutes)
- difficulty chewing
- difficulty breast feeding in infants
- coughing or gagging during meals
- excessive drooling or food/liquid coming out of the mouth or nose
- difficulty coordinating breathing with eating and drinking
- increased stuffiness during meals
- gurgly, hoarse, or breathy voice quality
- frequent spitting up or vomiting
- recurring pneumonia or respiratory infections
- less than normal weight gain or growth

Aspiration is unpleasant for the person experiencing it. It can cause all of the above but it can also be fatal.

Ways to minimize the risk of aspiration:

Positioning during meals/snacks

Appropriate positioning whilst eating and drinking, including when being tube fed or given fluids via tube.

Eg. A should be in her wheelchair or other appropriate seating with the back upright at a 90 degree angle when eating and drinking or being given fluids via gastrostomy.

Whilst eating orally

Tell her what's happening. Don't just start spooning food in her mouth. Encourage her to have an awareness of what she is eating and when it's going in to her mouth.

Eg. 'A I have some tomatoes here, do you want to try some.' Let her look at and smell the food before attempting to put the food in her mouth.

Enable the person to control the speed of the meal as far as possible. This may mean waiting for cues that they are ready for the next mouthful of food; looking at you, touching your arm, making their sound that says they are ready.

Make sure the person has finished chewing and swallowing before offering a drink. Alternate small bites with small sips. Be aware that it may take multiple swallows to clear the residue from one mouthful. Watch her breathing pattern as she eats. Wait and watch for her to swallow between mouthfuls.

Ensure there are minimal distractions during meal times. Don't expect the person to do other things whilst trying to chew and swallow food, i.e. Don't start putting on their coat whilst they are still chewing or similar.

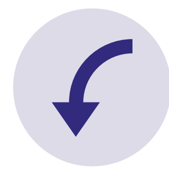
Modify the environment so it's calm in relation to noise, heat and light. Sensory distractions can compromise a person's ability to focus on what they are doing.

Talk to your SaLT about '**chin tuck**' and whether this could be useful to keep the person you care for safe from aspiration.

After food/fluids

Maintaining 45 degree positioning for 30 mins after food or fluids and ensure that this is written into any care plans at respite etc.

Eg. A should not be laid flat or tilted back in bed more than 45 degrees for 30 mins after food/fluids, including when on overnight feed/fluids.



An important note about reflux

Many people with Rett syndrome have reflux which often goes untreated. Suffering from reflux is generally unpleasant but it also increases the risk of aspiration. If you suspect your person with Rett has reflux, speak to your doctor about ways to treat this.

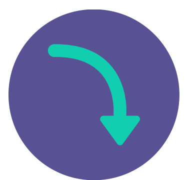
Many parents are put off checking whether their child has reflux as the PH probe which often assesses reflux can be unpleasant. People who do not have Rett are put on reflux medication every day after explaining their symptoms to their GP without ever being subjected to a PH probe.

Common signs of reflux are: [iii]

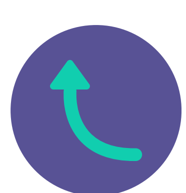
- regurgitation, sour smelling burps or vomiting
- dental erosion
- unexplained weight loss
- iron deficiency/anaemia
- food refusal and/or rumination
- recurrent lower respiratory tract infections
- behaviour problems (including agitation, self harm, screaming, restlessness for no apparent reason)

Some more things to be aware of...

Common anti-histamines can increase the risk of aspiration as they dry out the mouth and throat. Many people with Rett have common allergies and are prescribed allergy medication.



At Reverse Rett we often find that people with Rett syndrome have long term medication prescriptions which are not regularly reviewed which can lead to a person taking a medication several times a day, year round, which they do not need all the time and which could actually cause adverse reactions/events.



If your person with Rett is prescribed a common anti-histamine, speak to your doctor about appropriate times for them to take the medication. Is it every day, several times a day year round, or just when the person is symptomatic? Is it still necessary at all?

Brush her teeth.

Whether someone eats or drinks orally or not, it's very important to take care of their mouth and teeth.

Poor oral hygiene can cause a build up of bacteria in the mouth. People who have swallowing difficulties can breathe these bacteria into the lungs and then suffer from chest infections and serious respiratory difficulties.

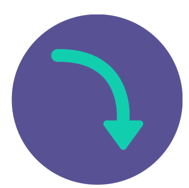
An electric toothbrush and some nice tasting toothpaste can make the experience quicker and more effective.

Always tell her what you are going to do before you start trying to brush her teeth. If it's difficult just do for a few seconds and build up over time. Gently clear her mouth of any food lying in the pockets at the side and front of her mouth. It's safe for her and no biting.

Give her a treat and praise her after you have brushed her teeth so she starts to be more accepting of the experience. Doesn't have to be food/snacks. Can be a favourite song lined up or tv show.

Videofluroscopy/barium swallow

There is an objective measure for whether someone is aspirating or not, called a Videofluroscopy. Videofluoroscopy is useful for the diagnosis of aspiration and to assess the need for texture modification.



During a videofluroscopy, a person is given food and or drink coated with barium to enable it to be visible on an xray. The person is then x-rayed whilst eating and drinking and the xray is observed by SALT and/or appropriate medical professionals.

A GP or Community Paediatrician can make a referral for Videofluroscopy.

A word about silent aspiration

Symptoms of aspiration usually appear after eating, drinking, vomiting, or an episode of heartburn but aspiration can also be silent.

Overt aspiration will usually cause sudden, noticeable symptoms such as coughing, wheezing, or a hoarse voice.

Silent aspiration tends to occur in people with impaired senses. Silent aspiration is when there is no coughing or clearing. Sometimes people aren't even aware that fluids or stomach contents have entered their lungs.

Some things to look for: red watery eyes or colour changes to the skin around the eyes, drooling or changes in the sound of someone's breathing, or their voice. Younger children often splay hands open also.

In summary

Thinking about aspiration is worrying. However, a recent paper by Tarquinio et al., focused on 'The Changing Face of Survival in Rett syndrome,' presents things differently. This paper concludes, 'Survival into the fifth decade is typical in Rett syndrome, and death due to extreme frailty has become rare.'

Whilst papers which largely focus on morbidity are not for everyone, the take home message from this paper is clear; **many risk factors for early death in Rett syndrome, including aspiration, are modifiable.** That means we can do something about it.

Further reading

- **Problems Swallowing? Resources for healthcare staff outlines the issues facing adults with learning disabilities who have dysphagia and includes support materials that can provide practical help for these people**
- **Public Health England, Guidance on Dysphagia/Swallowing Difficulties**
- **Gastro-intestinal disorders in Rett syndrome Checklist for clinicians on assessment and management**
- **Growth and Nutrition in Rett Syndrome: Checklist for clinicians on assessment and management**

[i] Tarquinio DC, Hou W, Neul JL, et al., The changing face of survival in Rett Syndrome and MECP2-related disorders. Pediatric neurology. 2015;53(5):402-411. doi:10.1016/j.pediatrneurol.2015.06.003.

[ii]Heslop, P, Blair, PS, Fleming, PJ, Hoghton, MA, Marriott, AM & Russ, LS, 2013, 'Confidential Inquiry into premature deaths of people with learning disabilities (CIPOLD): Final report'. Norah Fry Research Centre

[iii] Baikie G1, Ravikumara M, Downs J, Naseem N, Wong K, Percy A, Lane J, Weiss B, Ellaway C, Bathgate K, Leonard H. Gastrointestinal dysmotility in Rett syndrome. J Pediatr Gastroenterol Nutr. 2014 Feb;58(2):237-44. doi: 10.1097/MPG.0000000000000200.

M 07596 139466
E enquiries@reverserett.org.uk

reverserett.org.uk

Registered charity in England & Wales, charity number 1136809 and in Scotland, charity number SC046735. A company limited by guarantee number registered in England & Wales, company number 07278507