



Syncope Trust And Reflex anoxic Seizures

# Reflex Anoxic Seizures (RAS)



Working together with individuals, families and medical professionals to offer support and information on syncope and reflex anoxic seizures

[www.stars.org.uk](http://www.stars.org.uk)

Registered Charity No. 1084898

# Glossary of terms

## **RAS**

Reflex Anoxic Seizures (RAS) is a term used for a fit which results from a brief stoppage of the heart through excessive activity of the vagus nerve

## **12-lead ECG**

12-lead Electrocardiogram (ECG) is used to record heart rhythms whilst in hospital

## **Heart Rhythm Monitor**

Heart Rhythm Monitors are used to record heart rhythms for up to a week whilst away from hospital

## **ILR**

Implantable Loop Recorder (ILR) is a small thin device inserted under the skin to record your heart's activity

## **Tilt Table Test**

A Tilt Table Test is an autonomic test used to induce an attack whilst connected to heart and blood pressure monitors

## **Contents**

**What is RAS?**

**What is the difference between RAS and breath holding?**

**How is RAS diagnosed?**

**What should I do if my child experiences an RAS attack?**

**Avoiding attacks in given situations**

**Managing a child with RAS**

**What are the treatment options for RAS?**

**Night terrors**

**What can be done to help?**

**Living with RAS**



# Reflex Anoxic Seizures (RAS)

*"Reflex Anoxic Seizures (RAS) was first clearly distinguished in 1964 ("type 2 hypoxic crises", Mulsby & Kellaway). The term is used for a particular fit which is neither epileptic nor due to cyanotic breath holding (prolonged expiratory apnoea), but which rather results from a brief stoppage of the heart through excessive activity of the vagus nerve. I have used the term Reflex Anoxic Seizures (RAS) to describe the convulsions which accompany this particular type of syncope. Syncope (sin-co-pee) is derived from the Greek word to cut off. SYNCOPE is the result of the temporary cutting off of the supply of oxygenated blood to the brain. The mechanism of the syncope in susceptible individuals includes reflex cardiac standstill (always reversible) commonly following a surprising bump to the head or elsewhere. Associated marked pallor of the skin has led to these attacks being called Pallid Syncope. In more recent times these attacks have been referred to as Reflex Asystolic Syncope."*

**Prof. J B P Stephenson MA. DM. FRCP. FRCP.  
(London & Glasgow) Hon. FRCPCH. DCH.  
Founder & STARS Patron**

This booklet is for families, friends and individuals who care for a child who has been diagnosed with Reflex Anoxic Seizures (RAS).

RAS occurs mainly in young children but can occur at any age. Unexpected stimulus such as pain, shock or fright can cause the heart and breathing to stop with RAS often being misdiagnosed as breath holding or, more seriously, as epilepsy.

It is important to remember that although RAS is frightening to witness it is not life threatening.



# What is Reflex Anoxic Seizures (RAS)?

These occur mainly in young children but can occur at any age. Any unexpected stimulus, such as pain, shock, or fright, causes the heart and breathing to stop. The eyes roll up into the head, the complexion becomes deathly white, often blue around the mouth and under the eyes. The jaw will clench and the body will stiffen, sometimes the arms and legs will jerk. After what seems like hours but is probably less than 30 seconds, the body relaxes, the heart starts beating (sometimes very slowly at first) and the sufferer is unconscious. One or two minutes later the person may regain consciousness but can appear to be unconscious for over an hour.

Upon recovery, the person may be very emotional and then fall into a deep sleep for two to three hours. They may look extremely pale with dark circles under the eyes. RAS attacks may occur several times per day/ week/ month. The attacks appear to come in batches.

Unfortunately, because of the symptoms, it is known that RAS is often misdiagnosed as temper tantrums, breath holding or as epilepsy. It is one of the aims of STARS to improve both professional and public awareness of RAS.

# What is the difference between RAS and breath holding?

Breath holding is different to RAS in that during breath holding the heart continues to beat normally, but the breathing stops temporarily. Breath holding is not done on purpose but is also a reflex.

It is normal when crying, or when very upset, to have some difficulty “catching your breath”. The child makes an expiratory loud cry (breathes out with the crying noise), then is silent before gasping breath in. Sometimes, especially in some toddlers, they get stuck in the expiration and lose consciousness at that point. Typically, a child may look distressed and go very blue during the silent phase, then collapse unconscious, either floppy or stiff. This can look just like RAS. Although there are treatments for RAS when it is severe and frequent, there are no treatments for breath holding. As with RAS, it is frightening to witness but is not in itself dangerous, the loss of consciousness (syncope) is brief and recovery complete.

*"...just to know that other parents are going through the same emotions and stresses you are makes a big difference"*

Kerry,  
Mum of a toddler



# How is RAS diagnosed?

A correct diagnosis can be obtained by a good description from a witness and establishing a trigger to at least some of the attacks. This can be aided by keeping a diary of events. Try to ensure that your chosen doctor is familiar with the condition. If you are able to video an attack this also helps towards a speedy, accurate diagnosis.

- **12-lead Electrocardiogram (ECG)** is done to check for any features on a resting ECG that might suggest a genetic, inherited or familial heart rhythm disorder, and for heart rhythm analysis.

The following tests are sometimes used to help doctors make a diagnosis:

- **Heart Rhythm Monitor** to record heart rhythms for up to a week whilst away from hospital.
- **Implantable Loop Recorder (ILR)** is used to monitor heart rhythms for months at a time if the episodes are less frequent than every two weeks. The device can remain in place for up to three years.
- **Tilt Table Test** is an autonomic test used to induce an attack whilst connected to heart and blood pressure monitors. Ideally, this test should be performed with other autonomic tests as these aid diagnosis and also help in treatment.

If the tilt test fails to trigger an attack, other triggers can be tried, e.g. ocular compression.

This involves the doctor pressing on the child's eyes for ten seconds. It is a strong trigger for RAS, is safe, but is quite painful, so is not done routinely.

# What should I do if my child experiences an RAS attack?

1. Stay calm! At least outwardly even if you are panicking within. There have been reports of children going into a second attack as they recover from the first attack because a parent was hysterical and the shock of that triggered a second attack.
2. Make sure the individual is lying in the recovery position in a safe environment ensuring nothing is blocking their airway. This can be on your lap, bed, floor or sofa. (Family and friends will feel better if you say 'recovery position' as it gives them something to focus on and reassures them that what they are doing is correct). The child normally starts to regain consciousness during that time.
3. Check to make sure nothing is in the mouth to choke on, being careful not to get fingers clamped in the jaws of the child.
4. Try to time the attack - it will be much shorter than you imagine (in most cases).
5. Talk reassuringly (it is known that the individual can sometimes hear but is unable to answer) and comfort them upon recovery.
6. Video if possible to show doctors if necessary.
7. When the child regains consciousness, allow them to sit up when they are ready and able. They may feel weak for some time, so leave them to lie down if they prefer. Reassure, cuddle and give lots of tender loving care, as they are often very emotional.
8. If an older child has been incontinent, do not make a fuss, and help them to change as quickly as possible. The older the patient the more embarrassing it is for them.
9. If they wish to sleep, allow it. They often sleep for 2-3 hours. Some however are just drowsy and dizzy or disorientated. Reassure and just be there for them. They often feel afraid and insecure.

10. Many sufferers report that noise is amplified after an attack - everyone appears to be racing around and shouting or being very loud. Keep the environment calm and quiet around the patient. They feel 'fragile'.
11. They can be very emotional for a few hours or days - be understanding and reassuring. Slowly encourage them to get back to normal but do not force the issue at first. Allow them time.
12. Some patients and parents report night terrors and pains in the legs a few days before, during or just after an attack - reassurance and awareness is vital. Massaging legs sometimes helps.
13. It is unlikely that they will actually remember the attack so if they don't want to talk about it do not force the issue.

Although it should not be necessary to call a doctor, if the person has had a particularly nasty bump, it may be wise to seek medical advice.





# Avoiding attacks in given situations

**Sports:** Allow a child to participate in gymnastics, karate, football, etc. Let the supervising adult(s) know about it, just in case. An example, is a girl suffering with severe RAS but has NEVER had an attack while at gymnastics. She has had many falls and bumps but she knows she is likely to hurt herself so when it happens it is not a shock. Equally, she has arrived home and just knocked her elbow on a door handle and passed out immediately.

**Swimming:** Encourage your child to swim but allow them to sit on the side of pool with feet in the water and gently wet the face, hands, arms, to get used to the temperature before jumping in. Again it takes away the shock element. One child with RAS jumped in a pool and had an attack because the water was so cold. Now she always sits on the side, gets used to the temperature, eases herself in and then once used to it she is fine jumping or even diving in.

**Vaccinations:** Warn a child that it will hurt (a little), reassure, explain what is about to happen as it happens – while the nurse or doctor injects. This reduces the chance of an attack brought on by the sudden pain of the needle. This has proved to help; not only does it stop an attack happening but also reassures the parent because they feel they are doing something to help the situation.

Remember the child is a normal healthy person and SHOULD NOT be excluded from any 'normal' activity; however a responsible adult should be made fully aware of the condition and the action to take if necessary.

In general, we encourage carers to allow a child to stand, watch and become accustomed, and gain confidence before participating in an activity whether that be a game, party or just entering a crowded room or new environment.



**Teachers, Carers & Dentists:** Inform all that the child has a problem coping with pain and sometimes passes out. Advise them to make the child safe and put them in the recovery position. You could give them the **RAS information sheet** which explains what to do.

**Triggers:** If you know a certain situation will or may bring on an attack in your child (e.g. eating a cold ice lolly, playing in snow, playground rides etc.) then avoid for a while until the child is slightly older and gradually re-introduce them.

*"As a child I did horse riding, swimming lessons, school trips, in fact everything a boy of my age should be allowed to do"*

Oliver, school boy



# Managing a child with RAS

1. Do not avoid disciplining your child because you are afraid of an attack. If the attack was triggered because you had shouted or disciplined, do not then allow the child to later continue to do whatever he was originally doing. RAS is not caused by bad behaviour but if not handled correctly it can lead to behaviour problems later. Do not give in to the child because of the RAS.
2. Do not stop your child doing anything! (Unless dangerous). Pre-warn the child of danger so that if an accident happens it is not such a shock. For example, falling off the bottom of a slide at the park. The child is perfectly capable of going on the slide...when he does it is not a shock. It may still hurt but it is not unexpected.
3. Do attempt to give the child an explanation of RAS and answer any questions they may have, appropriate to their age. STARS has a useful booklet for children called **Jack has RAS**.
4. Even with very young children, avoid discussing 'their problem' in their presence, it makes them feel 'odd', 'different' and can make them become withdrawn. Try to make light of it but be aware of their concerns. Siblings can often feel they are the cause of the attack. Reassure and explain to them.

*"We are sure she will have a happy and fulfilling university life, just as we did"*

Cathy, Mum of university student



# What are the treatment options for RAS?

Although as yet there is no cure for RAS, advances have been made with the use of:

**Pacemakers:** Cardiac pacing can be provided as a treatment to improve the quality of life for some individuals who are more severely affected with RAS. (See [STARS Pacemaker booklet](#) for further information)

**Glycopyrronium, Atropine Sulphate and Methonitrate** are inexpensive, safe and effective drugs that prevent most reflex anoxic seizures but can cause harm in overdose. A doctor may prescribe one of these in the short-term for young children having severe and frequent attacks that cause distress to the child and their parents.

**Side effects may include:**

- Dry eyes – eye irritation
- Blurred vision
- Dry mouth
- Excessive thirst
- Feeling hot and sweating
- Difficulty in passing urine
- Not enough known regarding long term effects
- Must be given daily whether attacks are occurring or not

**Iron supplements** may be beneficial. There is suggestive evidence that a shortage of body iron (iron deficiency) may allow episodes to be more frequent. Ask your doctor if iron supplements might be prescribed.

## Maintaining a balanced diet and hydration

in adults has been proven to reduce the frequency of attacks. Poor hydration, like anaemia, increases the risk of syncope (fainting). As yet it is not clear if this helps children with RAS, although some doctors recommend increasing fluid intake (water, milk, energy drinks NOT carbonated drinks or drinks containing caffeine).



# Night terrors

Night terrors are brief periods (about 10-20 minutes) of screaming/crying and distress in the early part of the night. These normally occur in toddlers up to the age of 4-5 years. Night terrors occur typically in the first part of the night – certainly the first half of the night. It is very difficult, if not impossible, to arouse a child from a night terror. The child may have their eyes open, but will 'stare straight through you' as if not seeing you. In fact, the child does not see you and will not remember anything about the night terror in the morning.

A night terror is a disorder of incomplete arousal from deep sleep. Most of the brain is shut down but certain motor (muscular movement) circuits are active when they should in fact, be quiet. The result is thrashing around, screaming/crying out, walking up and down, kicking, unintelligible speech and the child apparently terrified. The child will not respond to voice, touch or reassurance. It is known that children with RAS tend to have night terrors just prior, during or immediately after a batch of attacks.

Unfortunately, we have yet to discover why. Children often complain of unexplained pains in their legs and occasionally their chest.

## Important Facts

- The child will not remember the night terror in the morning
- Parents are advised not to even mention it to the child
- Attempting to wake the child during the night terror is rarely successful
- The child is NOT ill
- There are no known long term ill effects
- Usually, night terrors occur only once per night and not every night
- Changes in routine and overtiredness can make them worse



# What can be done to help?

- Stay calm during the night terror
- If the child is thrashing around and walking back and forth, speak reassuringly
- If possible, comfort and hold your child close until it passes
- Only attempt to restrain the child physically to prevent self-injury
- Place anything harmful out of reach.
- If necessary lock doors and windows
- Reassure siblings that the terrors will do no harm and will go away
- Your reaction, and those of the siblings, may upset the child who is having the night terror. Reassure the child
- Remember that the child will have no memory of the incident the next day and should not be made aware of the incident as this can cause distress

# Living with RAS

*"I just wanted to wrap her up in cotton wool, sit and play jigsaws all day, knowing that she was unlikely to have an attack if she didn't move.*

*Such a response to caring for a child with RAS is natural, however parents need to balance the understandable need to protect their child from harm against the long term problems this can cause. Excluding children from everyday activities and risks can stunt their normal emotional and psychological development. Discipline and limit setting is particularly hard for families of these children but remain essential for the child's healthy development."*

**Trudie Lobban, MBE**  
**Founder & Chief Executive STARS**

## Reflex Anoxic Seizures are also known as:

- Reflex Asystolic Syncope (RAS)
- Infantile Vaso/Vagal Syncope (IVS)
- Vaso-Vagal Syncope (VVS)
- Reflex Syncope (RS)
- Neurally Mediated Syncope (NMS)
- Neurocardiogenic Syncope (NCS)
- Pallid syncope
- Stephenson's syndrome
- Vagal cardio-inhibitory fainting fit
- White breath holding attacks

## They are not...

- Temper Tantrums
- Epilepsy
- Expiratory Apnoea
- Syncope (RES)
- Blue breath holding attacks
- Prolonged Expiratory Apnoea (PEA)
- Valsalva syncope
- Apnoea attacks due to stopping breathing
- Simple faints
- Pseudo-syncope
- Pseudo-seizures
- Other cardiac causes with arrhythmia (abnormal heart rhythm such as Long QT etc)
- Deliberate Behaviours



# STARS

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Please remember that this publication provides general guidelines only. Individuals should always discuss their condition with a healthcare professional.

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Registered Charity  
No. 1084898

© STARS Published March 2007,  
Reviewed March 2012,  
Reprinted September 2012,  
Planned Review Date March 2014



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of Health

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