REFLEX ANOXIC SEIZURES
(REFLEX ASYSTOLIC SYNCOPE)

“Working together with individuals, families and medical professionals to offer support and information on Syncope and Reflex Anoxic Seizures”

www.stars.org.uk
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Syncope Trust And Reflex anoxic Seizures Group was originally known as the Reflex Anoxic Seizure Information and Support Group. It was founded in March 1993 after Francesca Lobban was diagnosed as having RAS and with the full support of Professor Stephenson, Consultant Paediatric Neurologist, Royal Hospital for Sick Children, Glasgow.

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Please remember these are general guidelines and individuals should always discuss their condition with their own Doctor.
REFLEX ANOXIC SEIZURES

"Reflex Anoxic Seizures (RAS) were first clearly distinguished in 1964 ("type 2 hypoxic crises" of Maulsby & Kelloway). 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."

FOUNDER PATRON: Prof J B P Stephenson MA. DM. FRCP. FRCP. (London & Glasgow) Hon.FRCPCH. DCH.

REFLEX ANOXIC SEIZURES occur mainly in young children but can occur at any age. Any unexpected stimulus, such as pain, shock, fright, causes the heart and breathing to stop, the eyes to roll up into the head, the complexion to become deathly white, often blue around the mouth and under the eyes, the jaw to clench and the body to stiffen, sometimes the arms and legs 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 and looks 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 mis-diagnosed as temper tantrums, cyanotic breath holding (prolonged expiratory apnoea) or as epilepsy. It is, therefore, one of the aims of STARS to bring about both professional and public awareness of RAS.
Reflex Anoxic Seizures are also known as

Reflex asystolic syncope (RAS)
Vaso-vagal syncope (VVS)
Reflex syncope
Neurally mediated syncope (NMS)
Neurocardiogenic syncope (NCS)
Pallid syncope
Stephenson’s syndrome
Vagal cardio-inhibitory fainting fit
White breath-holding attacks

They are not...

Epilepsy
Blue breath-holding attacks
Valsalva syncope
Apnoea attacks due to stopping breathing
Prolonged expiratory apnoea (PEA)
Simple faints
Pseudo-syncope
Pseudo-seizures
Other cardiac causes with arrhythmia (abnormal heart rhythm such as Long QT etc)

DIAGNOSIS

A good description by a witness, establishing the trigger, keeping a diary of events and consulting a doctor who is fully aware of the condition and takes a detailed history will lead to a correct diagnosis. If you are able to video an attack this also helps towards a speedy, accurate diagnosis.

Routine tests (mainly to rule out any other underlying condition) may include

- ECG / EKG - wires placed on the chest to monitor the heart, and hopefully record an attack
- EEG - wires placed on the head to read the electrical output of the brain
- Event monitor worn in an attempt to record an attack occurring naturally
- Implantable Loop Recorder (ILR) to monitor and record events (see separate booklet for further information)
- Tilt Table Test to induce an attack whilst being monitored with ECG and EEG (see separate booklet for further information)

**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.

**WHAT TO DO IN BRIEF...**

Everyone who has responsibility for a person with RAS, must know what to do, should an attack occur:

1. Make sure the individual is lying flat in a safe environment ensuring nothing is blocking their airways.
2. Talk reassuringly (it is known that the individual can sometimes hear but is unable to answer) and comfort upon recovery.
3. 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**

1. Allow a child to stand, watch, survey 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.
2. Don’t stop your child doing anything (unless dangerous) but pre-warn of danger so if an accident happens it is not such a shock to the child.
i.e. falling off the bottom of a slide at the park. The child is perfectly capable of going on the slide but if he knows that he may fall off at the bottom then when he does it isn’t a shock - it may still hurt but is not unexpected.

3. Allow a child to participate in gymnastics/karate - again warn. Typical example of a girl suffering with severe RAS but has NEVER had an attack whilst at gymnastics. 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.

4. 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. (A 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/diving - whatever!)

5. Vaccinations - warn a child that it will hurt (a little), reassure, explain what is about to happen as it happens - whilst the nurse/doctor injects. This reduces the chance of an attack brought on by the sudden pain of the needle. This has proved to really help; not only does it stop an attack happening but also reassures mum because she feels she is actually doing something to help the situation.

6. Inform, in a matter of fact way, to all teachers/carers/babysitters that - “..... has a problem coping with pain and sometimes passes out - please make safe, put in recovery position, etc.” and give them the RAS leaflet which explains in plain English what to do and not to exclude child from sports, etc.

7. Stress the importance of informing those responsible for the child but not to over-react, and if they want confirmation that children with RAS should be allowed into mainstream school, etc. then contact STARS. (This takes away so much anxiety for parents and schools because someone else is left to explain RAS and can also provide informative literature).

8. If you know a certain situation will bring on an attack (i.e. eating a cold ice lolly) then avoid for a while until the child is slightly older and gradually re-introduce.
9. Maintaining a balanced diet and hydration in adults has proved to reduce the frequency of attacks. Poor hydration appears to be linked with 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).

10. 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.

**DURING & IMMEDIATELY AFTER 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 mother is hysterical and the shock of that has triggered a second attack.

2. Make sure the child is safe.

3. Lie flat whether on lap, bed, floor or sofa. Parents often feel better if you say ‘recovery position’ as it gives them something to focus on rather than just looking at the child, also the child will normally start to regain consciousness during that time.

4. Check to make sure nothing is in mouth to choke on, being careful not to get fingers clamped in jaws of the child!

5. Try to time the attack - it will be much shorter than you imagine (in most cases)

6. Video if possible to show doctors.

7. When the child regains consciousness allow them to sit up when they are ready and able. Reassure, cuddle and give lots of tender loving care, as they are often very emotional.

8. If an older child has been incontinent don’t make a fuss, and help them to change as quickly as possible. The older the sufferer 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 / 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 sufferer. They feel ‘fragile’.

11. The sufferer is often very emotional for hours/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. Sufferers and parents report night terrors and pains in 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.

**MANAGEMENT**

1. 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.

2. DON’T avoid disciplining your child because you are afraid of an attack. If the attack was triggered because you had shouted or disciplined, don’t then allow 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. Don’t give in to the child because of the RAS.

3. Do attempt to give the child an explanation of RAS and answer any questions they may have, appropriate to their age.

4. Siblings often feel they are the cause of the attack; reassure and explain to them. Explain their body is over-sensitive and it is beyond the sufferer’s control. If a sibling says ‘it isn’t fair’ agree and say ‘yes it isn’t fair that ..... has RAS’. Don’t blame the sibling for an attack; this can lead to resentment and anger.

5. If attacks continue after the age of 5 years be aware of any bullying from fellow classmates - children can be so unkind!

6. Families who have been diagnosed within a short period of time and have had contact with the support group normally cope better in their management and understanding of this condition.

7. Telephone the support group for reassurance and to know that you are not going through this alone.
8. FINALLY - if an attack happens - don’t blame yourself, don’t feel guilty, there are occasions when nothing could have avoided it happening.

**TREATMENT OPTIONS**

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

**PACEMAKER** Cardiac pacing as a treatment to improve the quality of life for some individuals with RAS. (See separate booklet for further information)

**ATROPINE.** Atropine sulphate / methonitrate is an inexpensive, safe, effective drug that prevents most reflex anoxic seizures BUT can cause harm in overdose. It is used 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
- Hot and sweaty
- Difficulty in passing urine
- Affects ability to learn
- Not enough known regarding long term effects
- Must be given daily whether attacks are occurring or not

**NIGHT TERRORS**

Night terrors are brief periods (about 10-20 minutes) of screaming/crying and distress in the middle of the night. This normally occurs 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 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, hold your child close until it passes
- Only attempt to restrain the child physically to prevent self-injury
- Place anything breakable 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
Starting school, toddler group or nursery not only is an anxious time for parents but for young children as well. This may be the first time the child has been separated from mummy. It may also be the first time when the child has to learn to share and be given instructions from a ‘stranger’. All children have a settling in period. Some settle far quicker than others, whereas some do not react to their new regular surroundings for some weeks, if at all.

Many children find the transition from the home environment to a new environment a challenging time. A child with a medical condition such as RAS can be particularly vulnerable. Every child’s RAS is unique to them just as every child is unique to their parent.

**THERE ARE WAYS THAT SCHOOL CAN HELP**

Many children find the transition from nursery to primary and primary to secondary a challenging time. A child with a medical condition can be particularly vulnerable.

1. Be aware of the child as a whole person. It is important that children with RAS have equal access to all areas of the curriculum if at all possible.
2. Set up a meeting as soon as possible with the parents/teachers/school nurse/classroom helpers so that all are fully aware and understand the condition. Write an individual Care Plan (draft copy available from STARS) and review it termly or by individual arrangement.
3. STARS has a video for hire showing attacks to help all staff understand and know what to expect should the child in their care experience a reflex anoxic seizure.
4. Decide with the child how to handle telling the other children/students. Maybe describing that the child likes to have a ‘little sleep’ when they hurt themselves and if that happens they should call the teacher immediately.
5. Openness about RAS prevents it from being used as a weapon for bullies.
6. It may help to ask mum or a named contact to accompany the child on school trips or to swimming lessons; this may reassure mum as well as the teacher when away from the school environment.
In every other aspect the child is a typical 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.

And remember... the older the child the less frequent the attacks, and the chances of having a reflex anoxic seizure whilst at school is highly unlikely.

**ADVICE FOR STARTING A NEW SCHOOL / CLUB / AFTER SCHOOL ACTIVITY**

This can be a really exciting time: new lessons, new teachers, new activities, new uniform AND new friends; however it can also be a worrying time.

★ Will you find your way around?
★ What will your new friends be like?
★ Will you cope with new subjects?

Most young people feel this way, however if you have any extra concerns because of your medical condition you may need to take a little extra care, just for the first few weeks of term.

★ Will you have a reflex anoxic seizure in front of these new people?
★ Will everyone know how to take care of you?
★ Will people treat you differently?

In secondary school you move around from class to class with different teachers. Make sure you have informed your Form Tutor of your condition and have given them a copy of this booklet. **STARS** can provide an abbreviated fact sheet.

**The following advice should be followed for all new secondary pupils.**

- Have early nights for the first few weeks of term
- Keep a diary of two or three things you like about school and two or three things you don’t like
- If you have a problem, talk to your Form Tutor, Head of Year, best friend and **DO** tell your parents
- Give a copy of your timetable to your parents
For pupils with syncope and reflex anoxic seizures you should also...

- Talk to your friends about the condition
- Reassure them that you are just the same as they are but that your body ‘shuts down’ unexpectedly after pain or shock
- Tell them it is unlikely that it will happen however if it does tell them how they can help

If you have any fears or worries please tell your parents or contact STARS on 01789 450564, or Freephone 0800 0286362. Help and advice will be given and if you wish you could be put in touch with another teenager to share your thoughts with.

**SO REMEMBER...**

1. Everyone starting the new school or club with you is feeling just the same

2. Don’t be afraid to ask for help – the teachers expect it in the first few weeks

3. Don’t get overtired. Talk about your worries

4. Don’t let syncope or RAS prevent you from taking part in any activities

5. **Complete your PERSONAL PLAN and hand to the appropriate person in charge**

6. **ENJOY** your new activity!
WHAT ARE THE AIMS OF STARS?

- To act as an information and support group
- To bring about public and professional awareness
- To gather more information and to aid research

WHAT DOES STARS OFFER?

- Information and reports on the various types of Syncope including Reflex Anoxic Seizures
- Videos showing natural attacks, ocular compression and media coverage
- Linking with similarly affected individuals and families
- Information leaflets and literature
- Newsletters, regional, national & international meetings
- Freephone 24 hour helpline
SUBSCRIPTIONS AND DONATIONS

Your subscriptions are what keep STARS up and running. Subscriptions are a minimum of £15 or $25 or €25 a year. It is vitally important that you remember to renew your subscription. Without your help, we would not be able to provide our essential service to the patients and carers of Syncope sufferers. Thank you.

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