Patient Information booklet

Reversible Cerebral Vasoconstriction Syndrome (RCVS)
What is reversible cerebral vasoconstriction syndrome (RCVS)?

Reversible cerebral vasoconstriction syndrome (RCVS) is a neurological disorder characterised by a sudden onset of severe headache associated with narrowing of blood pipes (intracranial vessels) that supply blood to the brain. On brain imaging the narrowing of the blood vessels can look similar to “string of beads”. In addition to headaches RCVS can also present as medical emergency with strokes (ischaemic strokes or bleed), seizure or as brain oedema (brain swelling). Usually the narrowing spontaneously resolves within 3 months hence called reversible vasoconstriction.

What are the symptoms of RCVS?

The most common symptom of RCVS is sudden, intense headache referred to as like a “thunderclap” headache. The headaches are either widespread or located at the back or top of the head: They are often associated with nausea and sensitivity to light. The headaches can come and go and it is not unusual for them to reoccur over a few days to weeks as recurrent thunderclap headaches.

Majority of patients recover following the headache. RCVS can be associated with other acute neurological symptoms such as seizures and stroke. Stroke related symptoms are as follows - changes in vision; difficulty in speaking (dysarthria or expressive aphasia) or understanding others when they speak (receptive aphasia), loss of balance (ataxia) sudden onset of weakness or loss of sensation on one side of the body.

If you develop any of these symptoms whilst in hospital, please alert a nurse or doctor immediately. If you have been discharged home and develop any of these symptoms then you will need call 999 or if able to attend your local accident and emergency department immediately.

What are the causes and risk factors for RCVS?

RCVS usually affects young and middle aged adults between the ages of 20 to 50.

It occurs in both men and woman but is more common in woman. Woman usually present in their 50’s and men in their 30’s

In younger woman, it can sometimes be associated with the changes that happen after giving birth (postpartum), during pregnancy or when starting or changing birth control pills (contraceptive pills).

A lot of people who get RCVS tend to have a history of migraines.
Risk factors (Precipitants) that may be related to RCVS include:

- Use of prescriptions drugs, for example antidepressants, medications to treat migraines, immune-suppressant medicines, drugs to prevent bleeding following childbirth and some anti-Parkinson’s medications.
- Use of common over the counter drugs such as nasal decongestants and nicotine patches, and supplements such as energy drinks that contain caffeine and ginseng.
- Use of illegal drugs for example, marijuana, cocaine, ecstasy, amphetamines and LSD (lysergic acid diethylamide)
- Use of alcohol
- Head trauma
- High calcium levels in the blood
- Certain tumours
- Blood transfusions
- Immunoglobulin (IVIG) treatment
- Autoimmune diseases
- Blood disorders

If you need to discuss more about these risk factors you should speak to your consultant neurologist

Triggers for RCVS

If you develop RCVS you may notice triggering factors. These include, orgasm, exercise, stressful or emotional situations, Valsalva manoeuvres (for example straining, coughing, sneezing), bathing and swimming. It is believed these factors may result in change of cerebral vessel tone.

How is RCVS diagnosed?

Most people diagnosed with RCVS at The Walton Centre are initially admitted for investigation of intracranial bleed such as haemorrhagic stroke or subarachnoid haemorrhage.

At that time you will usually undergo tests to find the cause of your bleed.

A CT angiogram which is a scan to view the vessels of the brain is usually the first investigation.
If this doesn’t find a cause, your doctor may ask for a formal invasive cerebral angiogram to review the blood vessels of the brain in more detail.
Some doctors prefer or add an MRI and MRA (MR angiogram) scan to show the brain and its blood vessels in more detail.
These scans are used to look for causes of the bleed and are used to see if there is narrowing of the arteries in the brain in a manner that may suggest it is RCVS.

If this is the case, you are likely to have your care transferred to a consultant neurologist who will take a detailed history, review brain imaging and perform physical examination. A consultant neurologist would provide the diagnosis of RCVS.

Following brain imaging sometimes further investigations may need to be performed. This includes blood tests and a lumbar puncture to rule out other causes of your symptoms that may cause similar narrowing of the blood vessels.

Many people who present with thunderclap headache or TIA have no abnormality on the first scans. If this is the case you may only get your diagnosis in the follow up outpatient department.

The initial scans can be normal in 25% or more of cases: The chances of a normal scan following thunderclap headache is even higher if it is taken in the A+E department.

**How is RCVS treated?**

Currently there is no proven treatment for RCVS although some treatments help.

**Medication:**
The use of calcium channel blockers, the most common being nimodipine can help reduce headache intensity. Nimodipine therapy is safe, well tolerated and improves thunderclap headache by up to 83%. The recommended dose to commence nimodipine is 30-60 mg every 4 hours. The duration of nimodipine treatment will be decided by your consultant neurologist. Most people may remain on this treatment for 2 to 3 weeks depending on their presentation, although the dose and time may be reduced and / or extended depending on your symptoms and the results of any follow up scans.

Side effects of Nimodipine are low blood pressure, flushing, changes in heart rate, headache, feeling sick and feeling too warm. Please read the leaflet accompanying the medicines given to you to take home from hospital. If these side effects occur, seek medical advice.

**Don’t take alcohol or grapefruit juice whilst taking Nimodipine.**
Once you have finished the course of Nimodipine, you should have your blood pressure checked by your GP.

Supportive treatment is provided by early identification and withdrawal of triggering substance (medication and/or illegal drugs) that may trigger your RCVS and headaches. You will be advised to rest and take regular pain relief.
For symptomatic treatment of headache with simple analgesics such as paracetamol and opioids is recommended and triptans should be avoided as they exacerbate or cause RCVS.

It is advised to avoid medications that can cause constriction of blood vessels such as serotonin reuptake inhibitor antidepressants, over-the-counter nasal decongestants, and illicit drugs due to potential risk of recurrence.

It is beneficial that you do not get dehydrated.

**Going home**

If you have been admitted because of a subarachnoid haemorrhage or stroke, once your medical team are happy that your symptoms are improving and you are on the right treatment they will let you go home.

Whilst most people recover very well, some may need to have input from the therapy team to improve their recovery.

The neurovascular nurse team will speak to you and advise you on managing your symptoms and support your recovery (details at the end of this booklet)

**Follow up**

The neurologist will follow you up as an outpatient.

If you have been admitted to the Walton Centre, the neurovascular nurse team will call you about 2 weeks after going home to support your self-care.

You may require follow up scans such as a CT angiogram or MR angiogram. This follow up scan confirms the diagnosis of RCVS as the narrowing is reversible and should resolve completely.

Most people will have the repeat scan done between 4 and 12 weeks. This varies depending on the neurologist treating you and your symptoms.

**You should ask your neurologist or specialist nurse for further clarification of your follow up.**

**What is the prognosis of RCVS?**

The thunderclap headaches typically resolve over days to weeks. Similarly, visual and neurological symptoms start to resolve around the same time as the headache. The constriction in the arteries varies between people but usually lasts between four to six weeks. By three months, most people’s scans are back to normal.

You will be expected to recover well and back to being independent in your daily activities. Only a small number require help such as managing finances but are able to able walk independent.
The small proportion of people with RCVS who have developed a stroke would require further follow up with neurologist, physiotherapist and rehabilitation.

Sadly, some people go on to have chronic migraine-like headaches or low mood.

**If you find this is you, you must tell your GP, neurologist or neurovascular nurse to ask for support such as counselling or psychology**

The recovery section in this booklet will help with your self-care and so please read it and enlist support

**Recovering from RCVS**

**Stroke like symptoms**
This can occur in any part of the body but is usually confined to one part or the whole side of the body following a thunderclap headache. You may need rehabilitation if your symptoms are severe. Although rare, some patients develop stroke like symptoms without headaches. These can range from a slight weakness to a complete lack of ability to move. It may be permanent but usually gets better.

**If you get any of these symptoms once you go home, you should go to your nearest accident and emergency department immediately and inform them you have recently been diagnosed with RCVS.**

**Seizures**
Some patients with RCVS present to hospital after a seizure or multiple seizures. You may have been given medication to prevent further seizures but these are not usually required long term as recurrent seizures from RCVS are rare.

If you had a subarachnoid haemorrhage from RCVS there is a small risk of having seizures in the first year afterwards. One seizure or fit does't mean you have epilepsy in this context If this is something you are worried about please discuss this with your consultant neurologist or specialist nurse.

**You and your family should have advice on managing seizures: please ask your doctor or specialist nurse.**

If you have had one or more seizure, you are required to inform the DVLA and cannot drive until you have been given permission.

**Sight**
Patients with RCVS may experience visual problems such as blurring, blind spots, black spots and double vision. If this occurs, you should tell your consultant neurologist or specialist nurse so that your vision can be checked.

**If this occurs when you go home you should contact your GP or A+E urgently.**

Produced by: Selina Monahan Neurovascular ACNS
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Produced: August 2020
Review date: August 2022
If you do experience double vision, you may find it more comfortable to wear an eye patch over one eye. It is worth noting that most visual changes, like the headaches, resolve with time.

**Communication Problems**
The processes in the brain that control communication (i.e. speaking, listening and understanding) may be disturbed following RCVS. If this happens, and is problematic, you will normally be referred for speech therapy. Sometimes, but not always, your speech and ability to understand improve with time.

**Memory**
It is not unusual to be left with poorer memory following RCVS. Your memories before the onset of RCVS will be less likely to be affected. This usually improves with time, but recovery can be frustrating as often your memory and concentration aren’t as good as before.

Try breaking down tasks into smaller steps and using a notice board or notebook to help you. Keeping your home organised and making lists helps. If this problem is affecting your day to day life, you may need to be referred for rehabilitation as an outpatient.

**Personality Change**
Anything going wrong in the brain can cause a change in character or personality. Character changes often settle down or are adapted to with time. They can be as a result of the stroke or bleed but could also be because of the psychological trauma of the diagnosis. If they are on-going and troublesome, you should be referred to a psychologist for an assessment and treatment plan to help you cope. Recovery from any brain injury can take up to 1 -2 years, this will not normally be done until you have been given time to recover (usually about 3-6 months after).

**Extreme tiredness**
During the early stages of recovery, bed rest is one of the most important things you can do to help treat RCVS. You will need to pace your activities and rest once you get home particularly if you are having a lot of visitors or your house is very busy. Even simple tasks such as a walk to the local shop may leave you feeling exhausted. This will improve with time but may not go completely. You will know if you have done too much as you will be exhausted and may experience headaches. This often happens early evening or the next day and means you have to rest!

**Pain Relief for headaches:**
Headaches is one of the most common symptoms patient have to deal with on a daily basis. Simple analgesics such as paracetamol or opioids (such as codeine phosphate or liquid oral morphine) are usually recommended. You should take these regularly to begin with if you need them. The thunderclap headaches typically start to decrease in intensity and frequency over a few days to weeks. Once the headaches start to ease; you must cut down your pain relief. You can do this gradually and should know that taking regular pain killers for too long may make headaches worse not better! Certain
types of pain killers can cause constipation so please take the laxatives if they are given to you and eat a diet that is high in fibre and keep well hydrated. Drinking 2 to 3 litres of water per day, regular meals, ensuring a good sleep pattern and in some instances avoidance of certain triggers (caffeine, alcohol, cheese etc.) can help reduce the frequency and severity of headaches. There are also some non-invasive treatment options such as menthol sticks to rub on the forehead which may help. Drinking water and using pain relief at the start of a headache may also help.

You should aim to be off all regular pain relief as early as possible after your symptoms subside.

Headaches are often worse if you have done too much that day or the day before. You may be more prone to get headaches which is common after RCVS.

**If your headaches persist and are troublesome once you have stopped all regular medication, you should see your GP or speak to your consultant neurologist or specialist nurse for specialist advice.**

**Pacing**
You must rest until your headaches have eased and then you must pace your activity for about 4 weeks. Take a daily rest as well as some gentle exercise. Build up activity as you feel able and try to go outside for regular walks. Exercise can be built up gently, but we don't advise ‘extreme’ exercise until you have been given permission from your consultant neurologist.

**Unusual sensations**
Some people experience unusual or strange sensations in their head following RCVS or a brain haemorrhage. We are not sure why this occurs, but do not worry about them and they should ease with time.

**Fear of recurrent RCVS**
This is a very common fear for all patients. Recurrence of an episode of RCVS is rare. If RCVS has occurred due to medications, it is advised to avoid these to prevent further recurrence as the risk is unknown. It is suggested there is a higher rate of recurrence in patients with sexual activity as a trigger for thunderclap headaches during the initial RCVS episode. RCVS is rare and unlikely to have recurrence if you follow your neurologist’s advice. It is important to learn a coping mechanism as this fear may prevent your recovery progressing. Remember, you will have a delayed scan to confirm the vasoconstriction has been resolved and you will have a follow up appointment. You will only be discharged from the Walton Centre when we think it safe to do so.

**Sensitivity to noise**
This is not uncommon following a brain haemorrhage or RCVS episode. Everyday noise such as television or background conversations can be just as difficult to cope with as loud noises. This usually settles down with time as you recover.

Recovery is dependent on how you feel. You will need to pace activities for the first month or more after going home because of tiredness and headaches. Feelings of
anger, frustration and sadness are not uncommon. This may be due to the condition but could just as well be due to the sudden life changing event and so will pass. If you are affected, you should talk to your GP. You may need to be referred to a psychologist in order that you are able to manage.

Enlisting the support of the “Brain Haemorrhage Support group” affiliated to the Walton Centre, “Stroke Association” or “Brain Charity” also affiliated to the Walton Centre may help in that they can offer emotional support and recovery advice.

Not everyone is affected this way; some see this as a new beginning; a time to re-evaluate their life following the trauma.

Using a diary from when you are in hospital and for the following weeks or months can be very useful. By charting good and bad days, you will be able to mark your progress; this will help a lot when you have ‘bad days’ and will mark how far you have come with recovery.

**Everyday Activities**

**Driving**
Following a RCVS episode, if you hold a driving licence, you are legally required to notify the Driver and Vehicle Licensing Agency (DVLA). You will not be able to drive until you have received DVLA approval and your doctor has confirmed you are recovered. If you drive without telling the DVLA, then your insurance becomes invalid. This is because driving involves many different cognitive and physical skills as well as multi-tasking, decision making and problem solving. Driving restrictions are enforced by the DVLA and each case has to be approved individually. You must speak to your neurologist.

**Flying**
Once all follow up imaging has been completed and you have seen the neurologist as an outpatient. You should be safe to fly.

**Drinking Alcohol**
Don’t drink alcohol until the Nimodipine course has completed and you have reduced the number of pain killers you take. However, most people find they are not able to tolerate large amounts of alcohol following a brain injury. The government suggests that woman do not drink more than 2-3 units each day and men 3-4 units a day. Information websites on alcohol awareness is printed at the back of this booklet. If alcohol has been linked to the RCVS episode, then you should avoid it and discuss with your neurologist.

**Smoking**
Smoking tobacco is not good for your health and it is advisable that you give up smoking completely. If you plan to quit, it is preferable that you aim to reduce the number and stop as it is recommended to avoid nicotine patches as they can trigger RCVS. A free help line number is printed at the end of this booklet.
Returning to work
You can return to work as soon as you feel able from 4-6 weeks after the thunderclap headaches subside. However, most people need about 3 months, to recover. (Occasionally longer if there have been significant problems) It is advisable to go back to work on a phased return.

This means you build up your working days and hours according to how you are feeling. A phased return is often graduated over 4 weeks or longer and your employer has an obligation to adhere to it.

The working life service at the Brain Charity will help you if you are having problems with returning to work. The number is printed at the back of the booklet.

Sexual Activity
RCVS can be triggered in some people following sexual activity. The risk of recurrence of RCVS is very low. Provided the diagnosis of RCVS is confirmed on repeat cerebral angiogram with reversibility of vasoconstriction it should be safe to resume sexual activity. For people who have suffered with stroke adjusting to changes such as sensory impairment, weakness, etc will need to be meet. Please discuss this with your neurologist.

Starting a family
It is advisable to avoid pregnancy for the first 6 months following RCVS. If you plan to get pregnant please discuss this with your neurologist for further advice.

Exercise
Most people can resume minor physical activities couple of months following a diagnosis of RCVS depending on how well they feel. You should take advice from your neurologist or physiotherapist. Avoid strenuous exercise for the first few months as fatigue can be significant symptom following RCVS.

Dental Treatment
Dental treatment is safe after RCVS. Inform your dentist about RCVS as they will need to be aware of this condition if they plan on prescribing any medications for you.

Family and friends
Enlist the support of your family and friends whilst you recover. They will be a great help with shopping, transport and support if you feel down. Family and friends support is helpful during the first 4 weeks after you go home and will be the biggest influence in your successful recovery.

Where can I meet other people, who are going through similar experiences?
There are details of the brain haemorrhage support group, stroke association and the brain charity their contact details are at the back of this booklet.

Will I be entitled to any benefits?
You may be entitled to sick pay from your employer, or you are likely to be able to claim
benefits appropriate to your specific situation. You should contact a charitable agency such as Citizens Advice Bureau, Welfare Rights or the Brain Charity to discuss individual circumstances and what you are entitled to.
Going home information

Name .................................
Consultant...........................................................................................................
Date of Sub-Arachnoid Haemorrhage/ RCVS .........................................................
Date and type of treatment......................................................................................

Follow up advice:
You must inform DVLA

Driving restrictions..............................................................

Build up activity as you are able:
You will benefit from pacing activity and resting each day until you feel more recovered

Working.................................................................

See your GP for blood pressure and cholesterol check when the nimodipine stops.

Other :

Medications:

Nimodipine to stop .................................

Pain relief (aim to be off all regular pain relief in a month: if you can’t reduce pain killers for headache you should let the neurovascular nurses know as more specialist input may be required

Appointments:
Phone call to check on your wellbeing ..............................................

Follow up scan as per neurologist ......................................................

Follow up clinic appointment with neurologist ..............................

Produced by: Selina Monahan Neurovascular ACNS
Reviewed by: Dr R Menon Consultant Neurologist and Cathy Stoneley Neurovascular ANP
Produced: August 2020
Review date: August 2022
Walton Centre Contact Details

For any emergency please attend your local Accident and Emergency Department or see your G.P.

Patient Experience Team: 0151 556 3090
E-mail: PatientExperienceTeam@thewaltoncentre.nhs.uk

Neurovascular Nurse Specialists for practical advice: 0151 556 3325
or through hospital switch
(calls cannot always be answered immediately)

Walton Centre switchboard - 0151 525 3611

Cairns Ward - 0151 529 5637 / 5638
Caton Ward - 0151 529 5628 / 5629
Chavasse Ward - 0151 529 5079
Dott Ward - 0151 529 5633 / 5634
Lipton Ward - 0151 529 8884 / 8738
Sherrington Ward - 0151 529 5641 / 5642
Intensive Care and High dependency unit - 0151 529 5772/ 5773

For information regarding appointments please contact your Consultant’s secretary.
Useful Contact details

NHS 111
Telephone advice if you need urgent medical help fast but it is not a 999 emergency (or you do not have a GP)
Website: http://www.nhs.uk and search ‘emergency and urgent care’

D.V.L.A. (Driver Vehicle Licensing Authority):
DVLA, Swansea, SA99 1BN.
Medical Enquiries: 0300 790 6806
Or you can email via https://www.gov.uk/subarachnoid-haemorrhage-and-driving

Brain Haemorrhage Support Group affiliated to Walton centre:
bhsupport@live.co.uk
Facebook or via Brain Charity

Brain Charity: 0151 298 2999. www.thebraincharity.org.uk

Stroke Association:
Stroke helpline: 0303 303 3100 Supporter care: 0300 330 0740
Website: www.stroke.org.uk

Department of Work and Pensions: 0345 606 0265
https://www.gov.uk/government/organisations/department-for-work-pensions
Advice regarding benefits or money problems after subarachnoid haemorrhage can be gained from the Brain Charity who have a stall in the outpatient department of the Walton Centre

NHS Free smoking helpline: 0300 123 1044
www.smokefree.nhs.uk

NHS alcohol help: 0300 123 1110
www.nhs.uk/Livewell/alcohol
ADVICE FOR CARING FOR PUNCTURE SITE TO GROIN/WRIST POST ANGIOGRAM.

If the problem is considered serious, help should be sought from your local accident and emergency department immediately. Otherwise, you should contact your G.P., NHS direct or visit your local walk in centre.

- Do not strain or lift anything greater than 10lb for 7 days following angiogram
- Drink plenty of water or juice 24 hours before and 24 hours after the angiogram to prepare the kidneys and flush the dye from your body (about 2 litres or 10 glasses)
- Do not drive or operate machinery for 24 hours following angiogram
- Do not do any physical exercise or sexual activity for 24 hours following the angiogram.
- Keep the dressing on, clean and dry for 24 hours following the angiogram
- After 24 hours the dressing can be removed and a shower (not bath) can be taken.
- Clean and inspect the site and wash with mild soap and water. Dry and recover with a plaster until it is healed completely
- A bath and swimming is allowed once the site is fully healed.
- Report to your local hospital urgently if the site bleeds and not stopped after 15 minutes of firm (but not excessive) manual pressure.
- Report to your doctor/ A+E if there is any swelling, change in colour (paler or darker) or change in sensation to the leg/arm.
- Report to a doctor if there are any signs of infection such as redness, pain, swelling or pus from the site.
- Report to a doctor urgently if there are signs of excessive new bruising (a small amount of bruising is expected from the procedure)
- Rarely people can get serious complications following an angiogram which may need further expert management including urgent surgery and so if you are unsure, you should always let someone know if you think that the puncture site isn’t as expected.