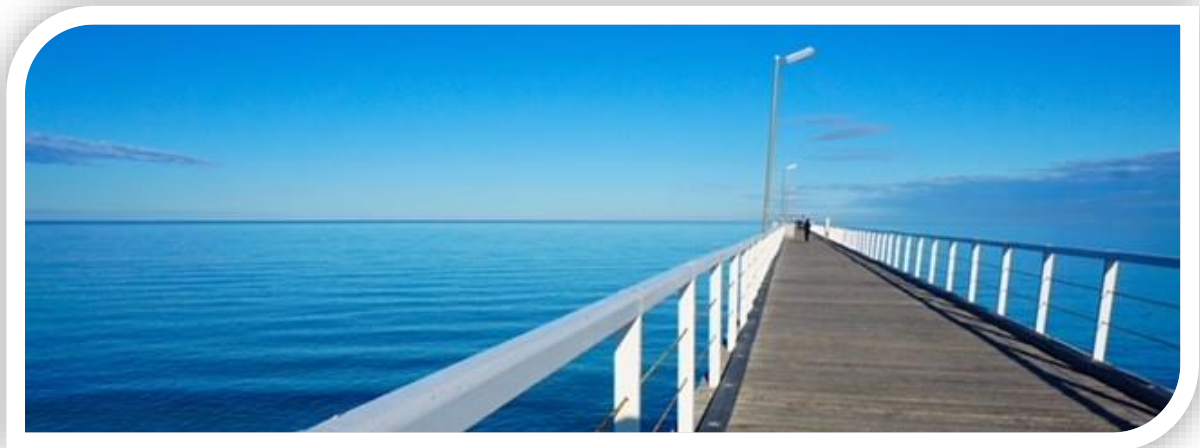


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Idiopathic Subglottic Stenosis

A Rough Guide for Beginners



Last updated: January 2018

Please join the Facebook Support group:

www.Facebook.com/groups/IdiopathicSubglotticStenosis

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What is idiopathic subglottic stenosis?

So you've just been diagnosed and it's all very overwhelming. Suddenly doctors are reeling off words you, your friends and family have never heard of, and when you search on the internet there is no or very little insight to be found. Well done for finding this document and hopefully our Facebook support group. We understand where you are coming from and this document will help.

Back to the question in hand. What is idiopathic subglottic stenosis (iSGS from hereon).

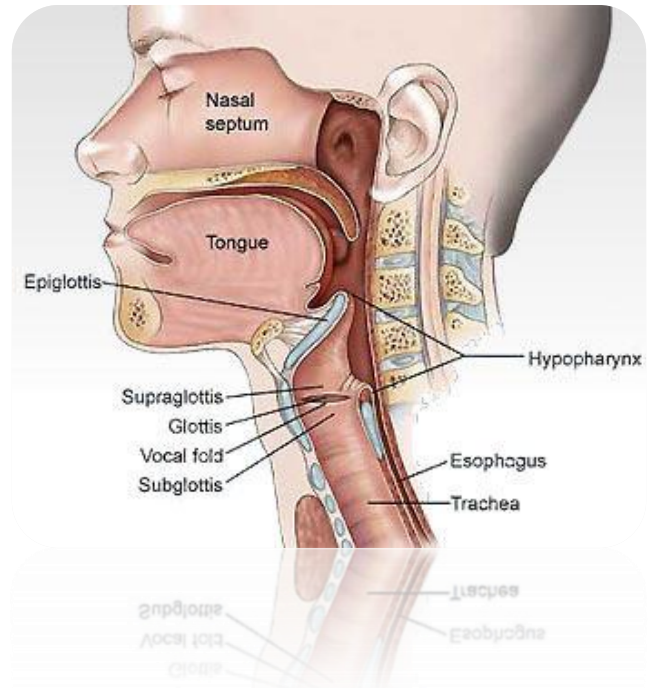
Getting down to basics;

- Idiopathic – is a fancy way of saying 'no known cause'
- Subglottic – is the part of the larynx below the vocal chords (glottis). It connects to your trachea ('wind pipe'). You may have been told you have tracheal stenosis – this describes a stenosis is located lower down, within the trachea
- Stenosis – another fancy word for 'narrowing'

There are four types of stenosis, all with the same symptoms but different causes. It is important that your doctor rules out the other three types of stenosis before you are sure you are idiopathic.

This document will help you regardless of your stenosis type, and of course, even if your stenosis is not idiopathic, you are welcome in the group.

As a patient with this disease, you are going to have to get used to being in control of your treatment – make sure you are aware of the treatment options available, the tests your doctor should be doing and be your own advocate. If you are uncomfortable with your doctor's level of experience, please request someone else. Your airway is not to be experimented with.



The four types of airway stenosis



Idiopathic – Doctors suggest your stenosis is idiopathic if you have no history of any of the following:

- Significant laryngotracheal injury
- Endotracheal intubation or tracheotomy within 2 years of your first symptoms
- Thyroid or major anterior neck surgery
- Exposure to radiation on your neck
- Caustic (eg chemical burn) or thermal (heat) injuries to the airway
- Vasculitis (a condition that involves inflammation of the blood vessels)
- Angiotensin converting enzyme (ACE) and antinuclear cytoplasmic antibody (ANCA)



Autoimmune - Patients with documented diagnosis of Wegener's (GPA), Relapsing Polychondritis (RPC), Systemic Lupus Erythematosus (SLE), Rheumatoid Arthritis (RA), Epidermolysis Bullosa (EB), Sarcoidosis, Amyloidosis or Mucous Membrane pemphigoid (MMP).



Polytrauma - Patients presenting with airway stenosis following documented traumatic injuries – particularly involving the trachea – such as breathing in chemicals or hot or burning air.



Iatrogenic - Patients that develop subglottic or tracheal stenosis following prolonged orotracheal tube ventilation (intensive care) or a tracheostomy – either immediately or within 2 years of intubation.

What are the common symptoms of iSGS?

We all share some common symptoms:

Stridor – this is what we often will describe as a wheeze, the sound when we breathe. It will often be silent most of the time, but get worse under exertion or in stressful situations. When your stenosis is particularly narrow, you may hear your stridor all the time.

Officially, a stridor is described as noisy breathing due to narrowing (stenosis) of the airway at or below the larynx.

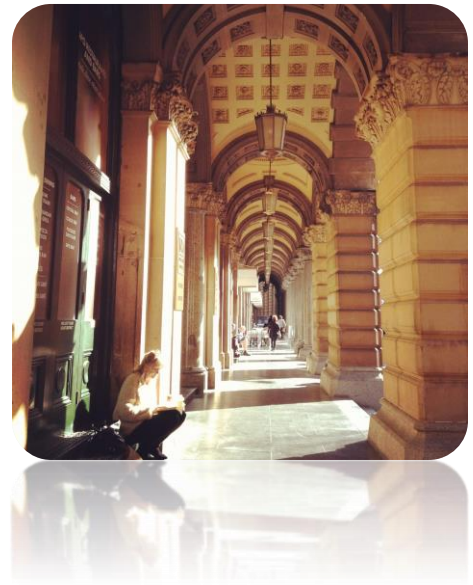
Often we as sufferers stop noticing this sound ourselves, and it is friends, relatives and work colleagues who might mention it to us. We often describe this as Darth Vader breathing!

Coughing – early on in your condition the coughing might not be too bad, but it tends to get worse over time. This is because the little hairs (cilia) which usually line your respiratory tract are absent where the scarring is. These hairs usually help move mucus smoothly up and down your airway. Where they are missing, you will need to cough the mucus past. The coughing will often get worse when you're talking lots, exercising or in stressful situations.

Breathlessness – it will be a struggle to breathe, talk, laugh and don't even think about singing! The more constricted your airway, the more challenging it will be. This is often what causes doctors to misdiagnose us with asthma.

Mucus – you will see many complaints about mucus on our discussion board. The fact is we don't have any more mucus than the average person; rather it becomes harder to shift because of the scar tissue. You may also find that you're more prone to getting minor bacterial infections within the mucus as it sits behind the scarring and thickens up. You'll have to cough hard to shift this and it might come up as a 'plug' – often thick white, yellow or green. If you're concerned, see a doctor, but mostly this clears up. Please see our tips for thinning and minimizing issues with mucus.

Inflammation – all this coughing and shifting of mucus can lead to inflammation. If it gets really bad (it can lead to a vicious circle of coughing and more inflammation) you may end up taking a short course of steroids. If you find you are prone to inflammation (your doctor may mention this after a scope), definitely look into our anti-inflammation tips which may help you avoid medication. Steroids have their own side effects, such as increased appetite (leading to weight gain), moon face (your face looks puffy and round), and with long term use, decreased calcium leading to brittle bones. Not everyone experiences these side effects, but be aware.



If you are like most of us, you were misdiagnosed for some months with asthma, bronchitis or panic attacks. You are most likely the first patient with this condition your local doctor has ever met. This means it is up to you to educate them!

Who gets iSGS?

Idiopathic subglottic stenosis is very rare – less than one in 200,000 people get this disease (the definition of 'rare') – it is in fact estimated to only impact one in half a million.

If you are female, then you are one of many, as this affects 98% females. If you are male then you are truly special and part of the 2%.

Many have not yet been diagnosed – as you will likely have found yourself, doctors often tell patients they have asthma or bronchitis, more common conditions.

People are often afraid of passing this on to their children – that is very unlikely. To date, with around 2500 members, less than 1% of the support group has a daughter or parent with iSGS, suggesting this is not something you should be too worried about.

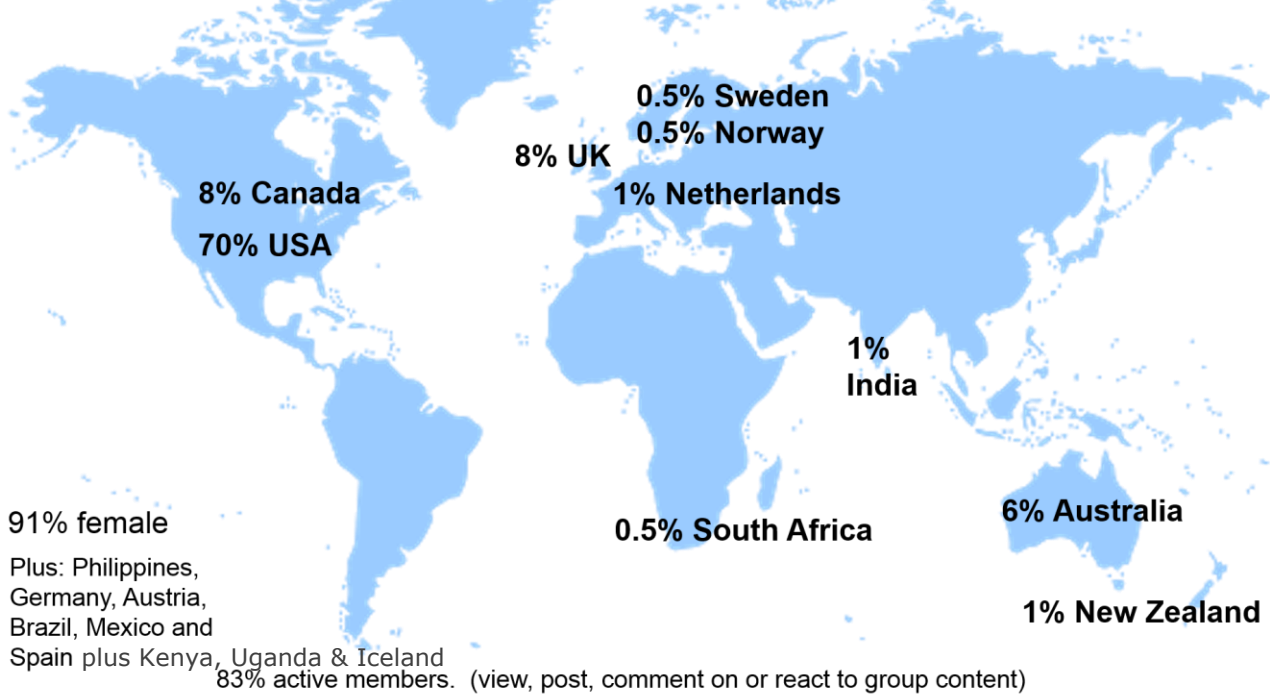
Eight in ten patients are diagnosed between 20 and 50 years of age, with a peak in the 31-40 age bracket. iSGS are no different from the rest of the population in terms of other health conditions (heart, thyroid, cholesterol etc). There seems to be a link with being white or at least some European ancestry.

One theory is that there is a hormonal link, but that has not been proven or disproven.

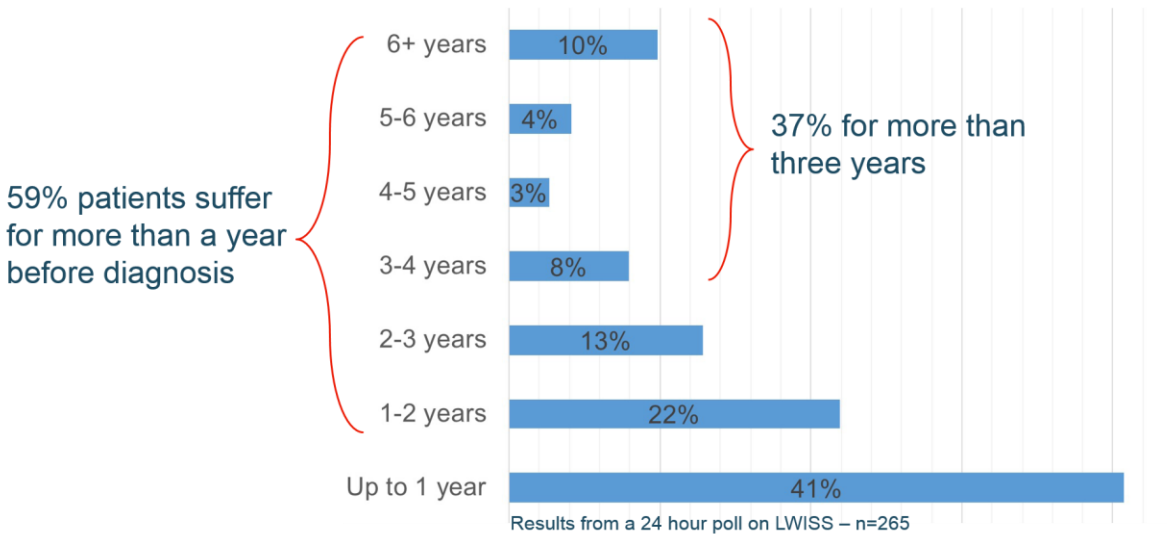


Here's some information about the Facebook Group members as of September 2017.

Key facts: Living With Idiopathic Subglottic Stenosis – the Facebook Group



Time from first symptom to diagnosis of iSGS



Debunking the myths about iSGS

There are so many theories about iSGS and its causes and effects. Sometimes inexperienced doctors are the ones telling you this. Let's nip them in the bud:

"iSGS is caused by reflux" – I have lost count of the number of times people have claimed this in the group. No. Your iSGS is NOT caused by reflux. Think about it. About 60% of the world's population has reflux – males as much as females – and yet iSGS only (well 98% of the time) affects women. Reflux is not the cause. It could be an irritant however. If you think this is the case, talk to your doctor about some reflux medication, and ask your dentist to check your teeth for any signs of acid erosion. Don't just take reflux medication 'just in case' – it causes its own issues such as brittle bones, kidney issues and more (and we really don't need any more health problems, do we?!)



"People with iSGS have more mucus than 'normal people'" – the fact is we have exactly the same amount of mucus as everyone else. The difference is that the cilia (little hairs in our trachea) which normally move the mucus up and down our airway with ease have been interrupted by the scarring. This means we have to cough it past the scar. Sometimes mucus can build up and thicken behind the scar and form a mucus plug. This can be quite scary as it can block your narrow airway until you cough it out. Check out our tips to help avoid this, and ensure you **have regular dilations** to avoid the potential of a plug you cannot shift.

"I cannot exercise because I have iSGS" Many of us make ourselves continue to exercise in spite of iSGS. You should not stop exercising – rather it will be beneficial if you keep as fit as possible. So what if you cough a little and sound like Darth Vader? The fitter you are, the better you will be able to cope with this disease.

Many people with this condition run, walk, or go to the gym several times a week despite a 5 or 6mm airway – just pick activities which are lower impact. We are superwomen (and rare supermen) – our bodies CAN cope with exercise.

There are plenty of alternatives out there if you look – these days you can get electric bikes which give you some exercise while helping you on the hills, try kayaking with a friend or your significant other in a double so they can help when you run out of steam...just don't give up moving because of iSGS.

We suggest you start exercising as soon as you're breathing well after a dilation and keep going as long as you can. Just do what you can within your limits and any other conditions you have.

"I am overweight because of iSGS" – again, most of the time this is an excuse. Most of us are overweight because we eat too much of the wrong type of food. Again, this is terrible for your overall health – you are putting stress on all your organs, and your iSGS is not helping. Work hard to reduce weight naturally – cut back on sugar and processed foods and replace with fresh vegetables. Monitor your calorie intake using apps or websites such as My Fitness Pal and become an overall healthier person. A recent study of iSGS patients showed that 58% are overweight or obese (sadly in line with the normal levels in most western countries). This needs to change...why not be part of it?

"iSGS is caused by being pregnant or taking hormones" – this cannot be totally debunked, but given that around one in five patients with iSGS has never been pregnant, that suggests it is not the only reason. Around one in four experiences worsening of breathing around the time of their period. One in ten iSGS patients has never taken any hormones (eg the pill). We also have males diagnosed with iSGS who definitely have not been on the pill or pregnant.

What tests should my doctor be doing?

There is not a definitive list of tests, but doctors in the USA suggest the following blood tests would be a great start to rule out known causes of scarring in the airway. Make sure you know what your doctor is doing or has done. Some of these might need to be done more than once.

- ESR (test for acute inflammation)
- CRP (test for acute inflammation)
- Rheumatoid factor (test for autoimmune disease)
- ANA (test for autoimmune disease)
- C-ANCA (test for Wegener's granulomatosis)
- Test for bacterial infections – in particular, Mycobacteria

Your doctor should also take a biopsy while under anaesthetic which will be tested.

It is strongly recommended that these blood tests are repeated each time you have an airway review or treatment (at least annually) as results can change many years later.

It is also recommended that your doctor take a biopsy every time a dilation is performed.



What medical interventions are available?

The following pages describe the various surgical approaches in more detail, however here's a summary of the surgical, medical and adjunct therapies you may come across on your treatment path:

Surgical procedures	Medical therapies	Complementary therapies
<p>ENDOSCOPIC (Using a flexible tube, inserted via the nose or mouth)</p> <ul style="list-style-type: none"> ○ Endoscopic tracheal dilation (with or without cuts or laser, with or without topical or injected corticosteroid, with or without Mitomycin-C) ○ Endoscopic resection with laser and medical therapies (e.g., antibacterial, inhaled corticosteroids, antireflux) ○ Endoscopic tracheal reconstruction (Maddern technique) <p>OPEN ANTERIOR NECK SURGERY</p> <ul style="list-style-type: none"> ○ Cricotracheal resection ○ Laryngotracheoplasty: <ul style="list-style-type: none"> ▪ Cartilage tracheoplasty ▪ Slide tracheoplasty ○ R-E-A-C-H-E-R (Retrograde, endoscopically-assisted cricoid hypertrophic epithelium resection) ○ Tracheostomy (rare) 	<p>ANTIBIOTICS to tackle bacteria which may be causing inflammation</p> <ul style="list-style-type: none"> ○ Bactrim, a sulpha drug ○ Azithromycin ○ Erythromycin, if allergic to sulpha drugs <p>CORTICOSTEROIDS to inhibit fibroblast formation and reduce inflammation</p> <ul style="list-style-type: none"> ○ Oral tablet (e.g., prednisone) ○ Oral inhalation (e.g., nebulized albuterol, budesonide, eg., pulmicort) ○ Injected (eg, triamcinolone, i.e., kenalog, between dilations in-office or "awake") <p>PROTON-PUMP INHIBITORS (PPIs) to reduce gastric acid reflux (GERD) if this is an issue for you'</p>	<p>HYDRATION to keep mucus thin and slippery (not thick and sticky) and to keep vocal folds moist</p> <ul style="list-style-type: none"> ○ Internal hydration: <ul style="list-style-type: none"> ▪ Drink plenty of water ▪ Reduce caffeine and alcohol ○ External hydration: <ul style="list-style-type: none"> ▪ Steam inhalation ▪ Nebulisation with sodium chloride solution ▪ Warm air room humidification ▪ Nasal rinse ▪ Nasal spray <p>MUCOLYTIC THERAPY to help expel mucus</p> <p>Mucolytic medication:</p> <ul style="list-style-type: none"> ○ OTC expectorant cough syrup (eg, guaifenesin) ○ Throat lozenges with glycerin, pectin or slippery elm (not menthol or eucalyptus) ○ Amino acid (N-Acetyl Cysteine, NAC): <ul style="list-style-type: none"> ▪ OTC oral capsule ▪ Oral inhalation (nebulized NAC, ie, Mucomyst) <p>Airway Clearance Technique or Device:</p> <ul style="list-style-type: none"> ○ 'Huff cough or 'huffing exercise ○ Flutter valve, eg, Acapella <p>LIFESTYLE CHANGES</p> <ul style="list-style-type: none"> ○ Dietary change, anti-inflammatory diet ○ Weight loss ○ Vitamins, herbs ○ Probiotics ○ Enzymes (eg, serrapeptase, bromelain)
<p>Note: This information has been compiled from published journal articles, physicians' recommendations, and personal communications. It presents iSGS testing, therapy or monitoring options that patients may be following. It does <u>not</u> intend to endorse or repudiate any iSGS testing, therapy or monitoring options.</p>		

Tips to help reduce mucus and coughing

We all cough all the time. It's a fact of life with iSGS unfortunately. Here are some tips to help thin the mucus and make the coughing a little easier (and less frequent hopefully!).

Diet

There has been some success with changing to the 5-2 diet. You eat normally 5 days a week and on 2 non-consecutive days you fast. This means eating no more than 500 calories for women and 600 calories for men. In a nutshell, the fasting days allow your body to focus on something other than digesting food - such as healing and cell renewal. Weight loss is a happy adjunct too! It's very easy to do. The great thing is, people doing this diet have found they have less mucus and cough less - surely it's worth a try! Don't expect results after just one week though - after two or three you should be noticing improvements...

Read more here: <http://thefastdiet.co.uk/>

Food to eat

- Grapefruit
- Pineapple
- Garlic
- Broccoli
- Raw honey (eg a spoonful to soothe a cough or sore throat)
- A teaspoon of peanut butter
- Chicken Soup - When University of Nebraska researchers tested 13 brands, they found that all but one (chicken-flavoured ramen noodles) blocked the migration of inflammatory white cells—an important finding because cold symptoms are a response to the cells' accumulation in the bronchial tubes. The amino acid cysteine, released from chicken during cooking, chemically resembles the bronchitis drug acetylcysteine, which may explain the results. The soup's salty broth keeps mucus thin the same way cough medicines do. Added spices, such as garlic and onions, can increase soup's immune-boosting power.
- Olive oil
- Fatty fish – eg salmon, tuna, sardines, lake trout, sardines
- Apple cider vinegar – 2 teaspoons to a glass of warm water, drink twice daily. You can add honey to enhance the flavour.

Food to avoid

- Dairy products
- Wheat & gluten
- Soy
- Sugar
- Processed foods

All the above are likely to thicken mucus and make you cough more.

Habits to stop

- Smoking! (surely nobody smokes with iSGS????)

Drinks

- Plain old water and lots of it
- Tall glass of warm water (non iced) with 1/4 fresh lemon squeezed into it - drink slowly over 30 minutes or so
- Herbal teas with a slice of lemon in - drink slowly and frequently throughout the day

- Coltsfoot tea (though read up about it first and only drink for max of 4 weeks at a time)
- Soda water (it breaks up mucus so it's easier to cough up)
- Thyme (you can make tea with it) clears secretions. In Europe, there are thyme teas and extracts for bronchial symptoms
- Pineapple juice mix - 1 Cup fresh pineapple juice, 1/4 Cup fresh lemon juice, 1 piece fresh grated ginger (about 1 & 1/2 TBS), 1 TBS Raw Honey, 1/2 tsp cayenne pepper. Mix all ingredients in a blender or juicer or stir in a glass until well blended. Drink about a 1/4 cup 2 to 3 times per day.
- Lemon verbena is also effective at loosening up and removing mucus from the lungs and sinus passages

Vitamins & herbal remedies

- Echinacea
- Goldenseal (also known as Golden Seal)
- Slow release vitamin C/Ester C 2000g
- Thyme
- Serrapeptase – an enzyme from silk worms. They use this enzyme to break down their chrysalis when they change into a moth. There is some (unproven) theory that taking this will help humans break down scar tissue.

Other non-chemical solutions

- Nebuliser - with saline or cool boiled water - 5ml at a time as often as you need it
- Humidifier in heated or air conditioned environments
- Neti Pot with warm water and salt
- Saline nose spray (as often as you need)

Over the counter medicines (manmade solutions)

- Mucinex (USA) or Robitussin Chesty Cough Capsul or liquid. Vicks cough syrup
- Bisolvon Chesty tablets (Australian brand) - contains bromhexine hydrochloride 8mg (breaks down mucus to make it easier to clear)
- Diffiam (may have other names in different countries - this is the Australian brand name) Anti-Inflammatory, Anti-Bacterial Lozenges - helps to reduce inflammation and coughing and mucus without the need for steroids (steroids to be avoided at all costs!)
- Nasacort (brand name in USA & UK) or Telnase (brand name Australia) nasal allergy spray - contains a steroid to reduce swelling in nasal passages. Can aid airway as well. Alternatively look for any nasal spray including triamcinolone (the active ingredient) - don't use long term (due to negative steroid side effects) or when you have a throat infection.

Tips to help reduce inflammation

Many of us have inflammation in our airway - this can be caused by excess coughing but can be eased. There are several natural options to try before going down the steroid route (which may be necessary short term, but to be avoided if possible!)

Food

- Berries - such as blueberries, raspberries, and strawberries
- Beetroot
- Apples
- Cherries
- Fresh ginger - try it in tea or stir fries
- Lemon - half a slice in tea, half a slice in water
- Eat fresh food you make yourself - avoid processed foods and high sugar
- Don't use vegetable oil in cooking - switch to extra virgin olive oil, grape seed, coconut or avocado oil
- Eat more fish rich in omega 3 - minimise red meat
- Snack on nuts and seeds - almonds, hazelnuts, walnuts, sunflower seeds in particular
- Fresh garlic - best uncooked!
- Herbs - especially mint (try chopped up in salad), basil, chili peppers, oregano, parsley, rosemary, thyme, and the wonderfully powerful turmeric.
- Dark chocolate (70%+)

Vitamins and minerals

- Omega 3 fatty acids - found in Cod liver oil and Fish oil
- Probiotics - friendly bacteria - you can buy supplements (usually refrigerated) or this is found in yoghurt (if you're not avoiding dairy!)

Beverages

- Water! Lots of it
- Minimise caffeine and alcohol - or at least match or double up with water when you consume these
- Green tea, Redbush tea
- Also, try the pineapple drink - see tips on reducing mucus

Over the counter (nonprescription) medicines

- Diffiam (may have other names in different countries - this is the Australian brand name) Anti-Inflammatory, Anti-Bacterial Lozenges - helps to reduce inflammation and coughing and mucus

What equipment will help me?

1. Nebulizer

We **highly recommend** you invest in a nebulizer to help with this condition. A nebulizer is a machine which basically converts liquid into a cloud which you breathe in through a mask. Most of the time all you will need to put into your nebulizer will be saline (available from chemists, online or may be provided by your doctor) or cool boiled fresh water (ie don't re-boil a kettle – empty it, boil a fresh batch of water and let it cool).

You may come across nebulisers when you are in hospital – they usually come straight out of the wall, are noisy and involve putting a mask over your face and breathing in. In the hospital, they may be mixed with pure oxygen or just the air in the room, plus saline (makes a gurgly noise as it is turned into clouds!)

How do I use it

Each model has its own instructions to follow, but generally, you fill around 10ml of saline or cool boiled water into a reservoir, reconnect the device, affix the mask over your mouth and nose, and turn on. Some models come with an alternative mouth piece if you don't need to be hands free, which allows you to just breathe straight in. Breathe normally, occasionally taking deep breaths to ensure you get the liquid deep down your trachea.

Why is it useful?

It helps put liquid directly into your trachea, soothing the airway and helping shift any mucus that is there, and breaking up any thicker lumps which might be causing a plug. It may make you cough, but this is good. You are only coughing because something needs to come out – have tissues on hand!

We cannot emphasise enough the importance of nebulising when you need to – in the 8+ years since the Facebook support group has been running, we have lost three members (that we know of) due to mucus plugs which blocked their airways. Nebulising can help you to avoid a life-threatening situation.

What types of nebulizer are there?

There are many on the market. Some that patients who are members of this group recommend are:

Omron MicroAir – portable nebulizer

Approx. \$170-\$200 online
Power adaptor adds approx. \$25



- Small and light – handbag sized
- Silent
- Portable – can run without mains power so can use in the car, watching tv, doing the housework or cooking
- Runs on batteries (last around 7 hours) or mains power (purchased separately – plug available for all countries)
- Some delicate parts so care needs to be taken when using and cleaning

Allersearch Portable Nebuliser

Approx. \$160-\$180 online



- Small and light – handbag sized
- Portable – can run without mains power so can use in the car, watching tv, doing the housework or cooking
- Takes AA batteries
- Noisy?
- No mains power option

Other brands Eg Allersearch Ventilair Max

Approx. \$210 online



- Reliable
- Hard to damage
- Runs with mains power
- Noisy – so only use when others not sleeping/watching tv
- Less portable due to size
- You are tied to the bed/chair where using as no battery option

2. Humidifier

A humidifier is a device you fill with water, turn on and it puts moisture back into your atmosphere. Particularly useful where you have air conditioning or heating on where the air is particularly dry.

How do I use it

Each model has its own instructions to follow, but generally, you fill a reservoir with water and turn on. You leave it somewhere in the room where you are working or sleeping.

Why is it useful?

It moistens the air you are breathing, making it less drying on your trachea and more soothing. Not as effective as a nebulizer but a less invasive way of easing the way.

What types of humidifier are there?

There are many on the market. You should easily be able to find them online – often second hand from parents who like to use them to help young children with croup.

Vicks Warm Steam Vaporizer

- It does what it says on the box
- It is hard to clean
- suitable for a mid-sized room
- Some report it trips power

Approx. \$50 online new



3. Pulse Oximeter

A device to check your oxygen saturation

Why is it useful?

It measures oxygen levels in your blood and allows you to monitor what is 'normal' versus 'abnormal' for you. Your oxygen levels should be 90% or above.

What types of oximeters are there?

There are many on the market. You should easily be able to find them online for around \$35

4. A medic alert bracelet

MedicAlert Foundation® is a non-profit, charitable, and membership-based organization dedicated to the wellbeing of others. Founded in 1956 by Dr. Marion and Chrissie Collins, the foundation's mission is to protect and save lives by serving as the global information link between members and emergency responders during medical emergencies and other times of need. As a non-profit organization, MedicAlert Foundation is governed by a volunteer board of directors, comprised of highly respected national leaders from the healthcare profession and business.

Once signed up, you can purchase a bracelet or necklace on which you can describe any conditions/allergies plus your MedicAlert number which medical professionals can call up for more information about your health. There are a number of designs.

<http://www.medicalert.org/everybody/difficult-airwayintubation-registry/for-patient/FAQ>

Your medical ID provides for a quick recognition of your medical conditions, allergies, medications, or treatment wishes; this leads to faster and more effective medical treatment.

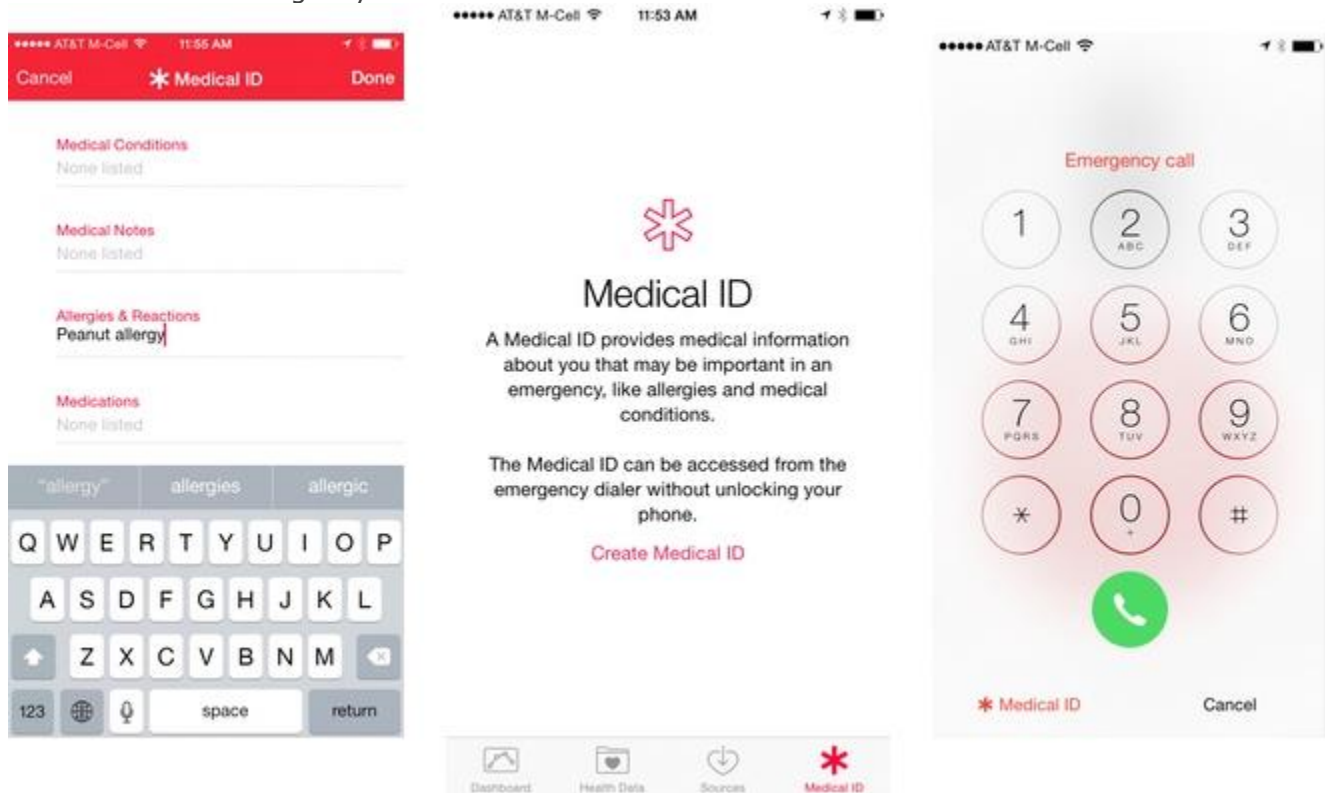
Medical ID bracelets reduce treatment errors which may result from not having a patient's health record during an emergency situation or upon hospital admission.

A medical ID speaks for you in the event of an emergency if you become unresponsive. First responders and medical personnel are trained to first look for medical identification jewellery in an emergency. Medical IDs will immediately alert emergency medical professionals to your critical health and personal information.

Suggested wording: Tracheal stenosis; difficult airway intubation; use fibre optics

5. iPhone users: *Medical ID

One of the lesser known — but potentially most important — features of Apple's mobile operating system is a digital "Medical ID," which can provide important personal health related information in the event of an emergency.



The Medical ID feature is built in to the Health application. Users can configure it by launching Health, tapping the Medical ID menu in the bottom right, and then choosing "Create Medical ID."

This is accessible even when the handset is locked – the medical professional can swipe the lock screen, tap "Emergency," and then view the information by tapping on *Medical ID.

A user's Medical ID can be configured with a custom picture and name, date of birth, list of medical conditions, notes, allergies, reactions, and medications. It also allows users to display an emergency contact with name, telephone number, and relationship.

The Medical ID also allows users to enter their blood type, height and weight, and whether they are an organ donor. And if someone changes their mind about having such information available from their lock screen, all the Medical ID information can be deleted via one button at the bottom of the editing page.

After the Medical ID has been created, users can always go back and make changes at any time through the Health app.

6. Peak Flow Meter

A peak flow meter is a portable, hand-held device that measures how well your lungs expel air. It will not improve how you breathe, rather it will help you understand whether you are stable or declining.

It measures your ability to push air out of your lungs in one fast blast by blowing hard through a mouthpiece (ie, the force of airflow through the bronchi and thus the degree of any airway obstruction). It gives you a numeric reading on a scale; peak flow readings are higher when the airway is open, and lower when the airway is narrowed (constricted).

Once daily use of a peak flow meter at approximately the same time of day helps you to measure day-to-day breathing trends and detect any airway narrowing before you may feel any symptoms. It can enable you to consult with your physician before your symptoms worsen.

How do I use my meter?

Take a deep breath, and then puff your air out sharply into the mouthpiece of the meter, making sure the little white marker starts at zero on your first puff. For your second puff, leave the white marker wherever it landed after your first puff. You might be able to move it a bit further. Try a third time, and wherever the marker is after your final puff, record this in the app (if you are part of the NOAAC research).

How often should I use my meter?

Ideally once a day.

Does it matter if it's not the same time each day?

Try for the same time of day, eg just before you go to bed. The app records time so the data analysts can choose to exclude times if there are clear differences at different times of the day, to avoid bias. You don't need to worry about this.

My numbers are lower/different to other peoples. Is this a problem?

Ignore the number you blow, we will all be very different, and concentrate on the long-term trend. Is it relatively stable over the month or is there a decline?

I have seen a chart of 'Predictive Normal Values', but my numbers are nowhere near these. Should I be worried?

Again, it is recommended you ignore the number you blow. For example, I have had an airway reconstruction, can climb and cycle very steep hills without stridor or wheezing and do high intensity training at the gym on a daily basis. My average number is 280. Per the Predictive Normal Values, I should be expelling between 450-500. ENT Doctors recommend you ignore this chart – it is not designed for iSGS patients.

My numbers are declining. What does this mean?

If it is in the short term, don't worry, but if you are consistently declining this may be a sign your airway is starting to close again.



This meter is showing lower numbers than my other meter. Why is that?

Peak Expiratory Flow (PEF) or peak flow readings vary widely between individuals and devices.

Where should I record my values?

If you are part of the NOAAC study, you can record PEF results using the Airflo app (<https://noaac.net/tools/>) or manually on a peak flow chart. Record the highest of three readings; do not reset the indicator between readings.



I just had a dilation or other clinical therapy, do I have to use my peak flow meter every day?

The last thing many of us would want to do after surgery is to blow hard on anything, so, no, give yourself a few days to recover. Resume use of your peak flow meter when it feels comfortable to do so.

Where do I get a Peak Flow Meter?

Peak flow meters are available for purchase at pharmacies, drug stores, and online retailers.

What are the benefits of recording my peak flow data?

You will learn what your airway is capable of when you are feeling well and when you are less well – you can also monitor how quickly you decline after a dilation. All evidence is essential to help your specialist offer advice about any potential next steps in your care.



7. Flutter valve device

This is a form of mucolytic therapy to help clear the airway of mucus.

A handheld flutter valve device clears mucus by combining positive expiratory pressure (PEP) therapy with airway vibrations.

Inside, the flutter valve, a small steel ball in a conical valve moves up and down as you exhale, vibrating your airway and making you exhale with resistance. This creates back (or positive) pressure that moves mucus from peripheral airways to larger central airways, so it can be coughed out more easily.

You breathe in and out regularly through the flutter valve five times, then in and out slowly five times, with exhalations lasting three times longer than inhalations. Alternate between regular and slow breaths, holding your breath two to three seconds after each breath. After five or six exhalations, try to cough. Repeat until no more mucus is expelled. This therapy takes 10 to 20 minutes.

*Acapella flutter valves pictured.



What is a dilation or dilatation surgery?

Once you have been diagnosed with iSGS your surgeon will most likely book you in for a dilation operation. While this does sound quite daunting at first (after all it may be your first ever visit to the theatre without enjoying the show!), hopefully we can help quell the nerves.

I've heard about laser dilation and balloon dilation. What is the difference between these?

Lasers can be used as either a cutting or ablative (evaporating) tool. It can cut through the scar to open it up or completely remove it. The bad part about the laser is that it uses extremely high heat to achieve its outcome that can cause further damage to the tissue. The balloon forces the scar open through a bunch of tears in the scar. The con with balloons is lack of precision – you can't control where the tears happen.

Ideally, we want to create an environment where the scar can't reform. This is done by having fresh wounds as far from one another as possible because fresh wounds close together will scar together.

Some doctors use a sickle knife to control the placement of the fresh cut wounds as far from one another as possible. A laser is not needed for this and saves the extra heat being applied to the tissue. Once the doctor has controlled the area of "tearing", they use the balloon to dilate the airway.

For thick scars, some doctors will use a laser to ablate the bulk of the scar making an effort to prevent extra heat from going to the normal tissue. This is done only at the thickest part and some scar is left surrounding it so there isn't a fresh circumferential wound, which can quickly reform a stenosis. They then use the balloon to further dilate.

How long does the dilation operation take?

It takes up to an hour in total. It may take longer if there are cuts made in the area with either a sickle knife or laser. Also, steroids or mitomycin C may be administered.

How will I breathe while I am being dilated?

The anaesthesiologist will intubate you with an airway tube while your doctor takes a look with the scope and gets set up. Your lungs are then hyper-inflated and the airway is pulled out while you are basically holding your breath and the balloon is inserted. While the balloon is in, they are carefully monitoring your blood oxygen levels to make sure they don't drop. You may have chest pain for a few days after the surgery- the hyperinflation of your lungs may cause this.

What is mitomycin C?

Looking it up online can be quite daunting when you find it is a drug used in cancer treatment (chemo). It does not mean you have cancer or are having chemotherapy. It is applied to your scarring at the end of the dilation in the hope that it will stop the scar cells from growing back.

There is no evidence that this works, but most patients do not experience any adverse side effects (you might be able to taste it as a bitter flavour when you cough once you wake up). Use of mitomycin C seems to be declining with most experienced surgeons.

How long will I be in hospital?

Most people are only in hospital for the day with some staying overnight. The reason for the overnight stay is as a precaution for possible swelling in the airway - particularly for people who do not live near the hospital.

How will I feel afterwards?

You will have a sore throat and feel sleepy from the anaesthesia but generally should feel good - within a couple of days you should definitely feel the benefit breathing wise.

Some people may have a husky voice for a few days post operation - at most for around 10 days - but this will go as any bruising subsides. In a few cases, people lose their voice for up to a month.

Your neck and back may feel stiff and sore from the positioning of you during surgery. The longer the surgery, the more prone you will be to being sore. You may also feel aching in your ribs from the hyperinflation of your lungs. All this dies down within a couple of days.

A saline nebuliser for 30 minutes a day for a few days post op can help soothe your airway.

What medication am I likely to be on afterwards?

Some doctors prescribe Tylenol 2 with Codeine (USA) or Panadene Forte (Paracetamol with Codeine - UK and Australia). This provides pain relief and suppresses any cough for the first few days.

Some doctors prescribe a steroid inhaler to use for a month post op.

Some patients are also prescribed an antibiotic (such as Bactrim) - they may be on this long term. The antibiotic is thought to help by controlling bacterial infections at the site of the scar. As doctors dilate, the tissue becomes damaged and dies off. It is thought that bacteria then colonize the wound and create further damage.

It is a relatively benign medication which would be OK to be on for a while (months to a couple of years) but it is not recommended to be on it long term for this disease.

How long before I should go back to work?

Usually recommend about 3 days away from work to rest post operation, with at least 2 days resting the voice post op. Some doctors recommend up to 5 days of no talking.

How long until I can resume normal activity or exercise?

Most patients are not given any advice on this - basically do what feels best. The day after your surgery you should be able to manage a short walk, but your throat will be sore so wouldn't advise running or anything too strenuous. Basically, go with gut feel. The anaesthetic will make you feel sluggish for a day or two, after that you should be able to go back. Best advice: listen to your body - if you don't feel like it, then don't do it! Don't over stress yourself and put back your recovery.

Will I only need one dilation?

Most patients will have a second dilation within two years, sometimes sooner. Some can cope more than two years between dilations.

What is a resection (resection and reconstruction) and how is it done?

A tracheal resection is a complex surgery that involves removing a scarred portion of the trachea and reconnecting the healthy ends.

Sometimes it can be called a resection and reconstruction. This is when the scarred portion is particularly long and a stent or piece of rib is inserted into the neck to replace the lost cartilage and prevent the neck from collapsing.

What is the success rate of this surgery?

Published articles suggest an 80-95% success rate. Recent research among iSGS patients finds that 73% of patients believe their resection was reasonably or very successful. Around one in five (18%) found their breathing more of a challenge after the operation – needing dilations again or another major surgery.

Can previous treatments 'mess up' your candidacy for resection?

The majority of skilled doctors will still be able to conduct a resection no matter how many dilations you have had. There may be other reasons you are rejected for a resection, however.

What exactly are the risks and the percentages of patients that experience any of these risks?

The main risk is restenosis. Next is damage to the nerves of the vocal cords. About 1% chance of bleeding or infection. Around 5% of patients with a very experienced surgeon will go on to restenose. 50% will have some change in voice (often deeper).

Around 90% of patients post resection will be fully functioning - the remaining 10% may have a weaker voice and inability to project. 2-3% will have permanent hoarseness if one of the nerves is damaged. Swelling after surgery is the biggest concern but only 1-2% will need a post-surgical trach temporarily.

How bad is the pain? How long are you on pain meds?

Talk to your doctor ahead of time about pain relief and muscle relaxers. Some say the position the chin stitch puts you in is the greatest pain. The hospital will manage your pain in the best way for you. Some report experiencing a pain level of maybe 7 out of 10 day one and two, 5 out of 10 day three and then maybe a 2 out of 10 for the final few days.

What is a chin stitch?

The chin stitch **was used in the past** to help keep the head in position but is now considered an old technique. It has been found to cause more problems than it stops. If your surgeon mentions they are planning to use a chin stitch (sometimes called a 'Grillo stitch' after the first doctor who used this technique) **please ask them to reconsider**. Do not be afraid to stand up for yourself on this matter, it could save your life.

Point your doctor towards the website created by experienced airway surgeon Dr. Paul Castellanos – airwaysurgeon.com – there he shows an alternative method which is far safer.

Did you have difficulty swallowing after surgery? How long before you can start to eat and drink?

Most are on a feeding tube for up to 5 days before being placed on a soft diet. You will likely be given medicine via IV to calm your stomach.

How long after surgery before they have you up and around?

You are encouraged to move around and are likely to be walking within the first two days.

I saw mention of a drain, what is that, where is it placed and for how long?

It comes out from your chest or beside your neck incision and it is in 3 or 4 days, it is more unattractive than painful.

Do you need to have an NG tube and catheter? If so, for how long?

You are likely to be given catheter. As long as you can walk they will let you take it out.

Are you on antibiotics or any other type of meds following surgery? If so, how long?

Some receive steroids to keep swelling down as a daily injection at the hospital and then taken orally on a tapering dose for 2 weeks. You may also be given acid reflux medication as a precaution.

Were you told not to speak for a period of time after surgery? If so, how long?

There are varying experiences – some are asked to speak right away, while others are put on voice rest for several weeks.

How long did it take for your voice to get as good as it was going to get?

This is not known for sure – some have to have voice coaching to strengthen their vocals and return to previous levels. Others will always have a weaker or raspy voice.

What is the recovery time and what can I do during this period?

Minimum of 6 weeks off, and return to work depends on each individual, as everyone feels and recuperates differently. No driving for 8 weeks

How bad is the scar?

The scar is about 3 inches in length.

Can also have a small 1 cm scar where the drain is located

What is The Maddern Technique (endoscopic tracheal reconstruction)?



How is The Maddern Technique different to a resection?

In a resection, the affected piece of trachea including the cartilage is removed via open surgery. With this reconstruction, the scarring is removed and a skin graft placed temporarily into the trachea for 2 weeks to encourage healthy cells to re-grow in place of the diseased tissue.

This operation is endoscopic rather than via open surgery meaning the neck is not cut open. This has several benefits including a minimal risk to the vocal chords and the nerves that control them. Recovery is much faster as there is not a physical cut in the neck nor a section removed.

Broadly what happens in the operation?

The scarred skin within your trachea is removed – just the skin, no cartilage. A piece of skin is grafted from your thigh or cheek and wrapped around a plastic tube (stent) and inserted orally. This is then stitched in place with two small stitches at the base of the tube. All that is visible on the outside of your neck are the stitches. Depending on where the stenosis is, a temporary tracheostomy is necessary too.

The stent is left in place for two weeks and then removed. One of the benefits for this method is that there is no scarring from the reconstruction, other than a small scar (about 1cm) from the tracheostomy if that was needed.

If I need a tracheostomy, how long is this for?

The tracheostomy is provided to allow you to breathe while your airway is swollen and the stent is in. You will have this for around 5-7 days. If you don't bleed too much and your airway can cope well with a size 12+ open stent then you are unlikely to have a tracheostomy.

When can I get up and walk around?

Usually the following morning after your operation you will at least be able to visit the bathroom (you are not given a catheter and the bed pan is not a fun experience!), and you will feel more and more like exploring as the days pass. Once you are off the feeding tube and IV you will be able to make more extensive journeys to the coffee shop and even outside (as long as you let the nurses know when to expect you back!).

When can I eat and talk normally?

Once a speech therapist has visited you and ensured you can swallow properly you should be able to have the feeding tube removed. This depends on when they make it to see you - any time between 1-3 days.

How long am I in hospital?

You are likely to be in hospital for around one to two weeks. You may then be allowed home under strict instructions to remain healthy, nebulise lots and not overdo things, before returning for a day surgery to have the stent removed. Depending on your recovery you might be in hospital a little longer – up to two weeks in total (or until the stent is removed).

A month later you will need to return for another day surgery to laser 'tidy up' the new skin and so your doctor can check out how it is healing. You may need another final laser 'tidy up' a month later if only 50% is done the first time. Some patients will also need a third and final tidy up.

Will I be in pain?

The hospital is good at managing any pain you have with regular pain medicine (sometimes too regular!) and pain is minimal. Most of the discomfort will be from any stress you have, so ideally learn to recognize this and use relaxation techniques to calm down.

How long until I feel better post op?

As soon as you are recovered from the general anaesthetic from having the stent removed you will be back to exercising and normal life. You should continue to nebulise daily to help with the healing process.

Some people may still have some difficulty with mucus – after the surgery you will need two 'tidy-ups'- these are to ensure the graft is all removed and your airway is smoothed.

The graft site on your leg or inner cheek will be the sorest point, so keep this covered and protected. It's important while in hospital to ensure the original dressing is left on and kept dry to aid the healing process. Tape a rectangle of plastic bag over the site when showering to protect it.

How long does it last? How will I feel a year later?

The first patient to have this operation is breathing well 6years later. It is a new operation so there are no long-term case studies or statistics, but most patients so far are doing well.

Like all surgeries for this disease there are no guarantees on long term success. There is no fix all surgery. This may work for you long term, or just for a few years.

What is the REACHER?

At the time of writing this technique is only practiced in the USA by Dr Robert Lorenz at Cleveland Clinic (www.clevelandclinic.org).

The REACHER stands for Retrograde Endoscopically Assisted Cricoid Hypertrophic Epithelium Resection. It is similar in many ways to the Madden procedure. Both remove the diseased lining of the cricoid, while preserving the outer cartilage. Essentially the differences are that the REACHER is done through an incision in the neck (transcervically) - which is closed at the time of the surgery, vs. the Madden which is done through the mouth (transorally).

Both the REACHER and Maddern Technique remove all the subglottic scarring, and replace it with skin graft from the thigh with the use of a temporary stent.

Compared with a resection, the recovery of both surgeries is much faster, and risk of complications greatly reduced.

There is some expectation that the Maddern procedure will become the standard with the REACHER reserved for patients with either hard to visualise subglottic areas through the laryngoscope, or else patients who cannot tolerate jet-ventilation (their oxygenation cannot be maintained throughout the endoscopic procedure).

What are in office/awake steroid injections

More and more frequently, iSGS patients are choosing to have a steroid injection in the doctor's surgery. Here's more information about that technique.

Why would I choose to have a steroid injection?

Inflammation is a key part of iSGS and often causes major issues with our breathing, even when the actual scarring is not too bad. Taking regular oral steroids can help, but have many negative side effects. A single injection directly at the site of the stenosis can have an immediate effect and can keep you breathing well for much longer – keeping the need for more major surgery at bay.

How often would I need an injection?

You are generally seen every three weeks for a course of 4-6 injections.

How long does the procedure take?

It takes 5 minutes.

Am I sedated or put under a general anaesthetic?

There is no general anaesthetic involved nor sedation. The area is first numbed with an injection of lidocaine, a local anaesthetic. This will cause you to cough, spreading the anaesthetic through the trachea and voice box.

After it is numbed, what will the doctor do?

The doctor will use a scope to inspect the area, then inject the steroid into the scar.

Does the doctor inject from inside or outside of my neck?

This depends on your airway and the ease of inserting the scope with needle. Given a choice, many suggest internal injections are less painful than external injections.

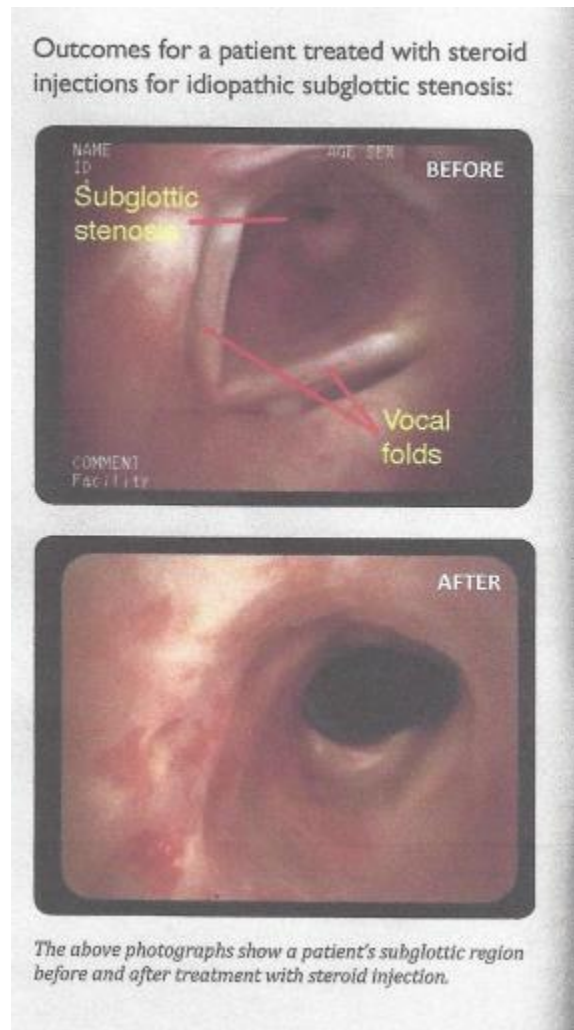
Will it affect my voice?

Some have a short-term huskiness while the vocal chords are numbed. This is temporary and usually eases within an hour of the procedure.

What difference would this make to my dilations?

Clinical data has shown that some patients can cope for long periods between dilations, with some having potentially permanent results. There is some suggestion (though at this early stage, not yet proven) that some patients would not need another dilation ever.

*Image source – Harvard Otolaryngology, Issue 10, Spring 2014



What is the Mayo Protocol?

The Mayo Protocol is a term you may hear from members of the 'Living With' Facebook Group. You might like to ask your doctor about trying this, or perhaps they will suggest it to you.

Several patients are following this regimen, often with good results.

The protocol involves taking three drugs:

- Bactrim – a sulpha based antibiotic – just small doses regularly. This potentially kills any bacteria in your away – patients with a stenosis tend to often have bacterial infections which build up behind their scarring, causing thickened mucus and the risk of plugs.
- A steroid inhaler – such as Qvar, Pulmicort or Flovent – one inhale per day. This reduces inflammation in your airway.
- Protein pump inhibitors/Acid reflux medicine – such as Prilosec or Nexium once a day. If you have reflux, then this can potentially reduce inflammation in your airway and prevent damage to the skin there from stomach acid.

What are the risks with this protocol?

As with all drugs, these can have side effects.

- Antibiotics: Some people can have negative side effects with taking antibiotics long term, such as thrush and stomach issues. Some people cannot tolerate sulpha based drugs, in which case an alternative is found.
- Steroid inhaler: This can cause oral thrush – you need to wash your mouth out and gargle with water after you take your breath. Some people can find the powder will coat their vocal chords (another reason to gargle), causing a husky or raspy voice
- Protein Pump Inhibitors/acid reflux medicine – if taken long term can potentially cause kidney problems and brittle bones

What are the benefits with this protocol?

Those patients who have had success while following this protocol have reported:

- Lasting longer between dilations
- Less inflammation, therefore improved breathing
- Less thickened mucus, therefore reduced coughing and improved breathing

There is no evidence that this stops the scarring, rather it tends to treat the added symptoms surrounding it.

I am considering major airway surgery. What should I ask my surgeon?

- How many resections/reconstructions have you done? How many on idiopathic patients? How many on non-idiopathic?
- What is your success rate? How do you define success?
- What can I do to protect my teeth from damage, if anything?
- Can I talk to any of your former patients?
- What is your major complication rate?
- What do you see as my personal risks?
- What happens during the operation? How long does it last?
- Is there a list of things to bring to the hospital?
- How long will I stay in the hospital?
- What can I expect when I wake up? (Lines, drains, masks, SCDs)
- What is the length of trachea removed?
- How bad is the post op pain? What kind of pain meds (Tylenol, muscle relaxers) and for how long?
- Will I need antibiotics? Steroids? For how long?
- Is swallowing difficult after surgery?
- Is there a drain? Where?
- When can I get up and walk? Will I need heparin shots?
- Will I need a catheter? When can I have the catheter removed?
- When will I be able to have ice chips? Solid food?
- What are the risks of: Restenosis? Nerve damage? Infection? Trach? Weak voice?
- Will I have a neck brace?
- How many days after discharge should we stay in [the city of surgery]?
- We have a long drive home. How many hours of driving per day?
- What restrictions will I have when I go home? For how long?
- Will I be able to sing afterwards? [Great question if you were never a singer!]
- Where and when will I go for follow up appointments?
- What restrictions will I have when I go home? For how long?
- How many weeks until I can drive?
- Any preoperative exercises recommended? Should I lose weight? Should I gain weight?

I'm heading into hospital – what should I take?

Going into hospital, even just for a day, is a daunting experience and if we can help make it easier with a checklist of things to take it will remove one more thing from our already busy minds (breath in, breath out!).

Going in for a day

- Book or iPad for killing time while you wait
- Lip balm as the air conditioning is quite dry (make sure it's clear as a tinted one will end up smeared all over your face - not a good look!)
- Phone with internet connection so you can keep your 'Living with idiopathic subglottic stenosis' group mates up dated and they can keep you company ;)
- Wear big knickers, as these will be your only clothes ;)
- (Apart from those you arrive and leave in of course!)
- Flip flops or slippers for walking around on cold tiled floors
- Any favourite snack that you can cope with eating rather than the hospital food once you wake up



Going in for a night

Same as going in for a day plus:

- Wash (toiletries) bag - definitely want to clean your teeth, possibly your own soap
- Tissues
- Night-time routine things - face creams and so on - help you try to get some sleep (it will be hard!)
- Night dress or pyjamas
- Laptop/tablet (if the hospital has free wifi!)

Going in longer term (eg a resection or reconstruction surgery)

Same as going in for a night plus

- Several pairs of underwear
- More extensive wash (toiletries) bag - shampoo, conditioner
- Yoga pants/loose trousers
- V-neck loose t-shirts
- Hair ties or clips to keep hair out of eyes
- Fleece blanket
- Load your iPad with movies and music - bring your headphones
- Ear plugs and sleep mask

- Bose Noise Cancelling ear buds – can be used to listen to movies and music, as well as just turning them on at night and popping them in your ears on silent as very effective ear plugs – highly recommended!
- Magazines (you can ask visitors to bring these too!)
- Any hobbies you can do while in bed (crosswords, knitting, crochet, drawing etc)
- A travel pillow to help support your head --- this was especially helpful for resection patients with the vampire stitches
- Sore muscle rub/balm. eg Icy/Hot.
- Extension lead with multiple power points for plugging in your laptop/iPad/phone/etc
- Netflix subscription! Great to watch a whole TV series while recovering
- Travel hair dryer - luxury!

Tips on surviving major airway surgery in hospital

If you are heading to hospital for a resection or reconstruction, then definitely take note of this checklist for survival.

Pre op be as healthy as possible. Lose weight if you have excess, eat healthily, do as much exercise as you can. Many can credit their overall recovery to this regime. Some do the 5:2 diet for a year before their op - this reduces your sugar levels, bad cholesterol and puts your body into repair mode as well as weight loss. Ensure your body is working at its most efficient to help repair the surgery site.

Focus on the end goal, what you want to achieve from the operation and what you want to do with that. Do not dwell on the procedures and how they may or may not make you feel. Never lose sight of what you want from your operation.

Try to recognize progress every day, however small it is. It will motivate and encourage you - eg - being allowed to use the bathroom rather than a bedpan, getting your feeding tube removed, being allowed to wash yourself, then shower...even taking fewer pain killers is progress...

Move and sit up - As soon as you can, get out of bed or at least sit up. This will help clear your chest and reduce coughing and risk of infection. If you can walk around the floor a few times this is even better. Get the blood moving and help uplift your spirits.

Wash - As soon as you're allowed to shower or at least wash yourself then do. All about empowerment and besides, warm water feels amazing! If you can wash your hair, even better - it's so healing

Establish your own routines. Again about gaining a bit of your own power rather than being solely at the mercy of the hospital system eg get dressed during the day and change into your gown in the evenings, use your own eye cream, lip balm and so on before you sleep.

Talk - not talking is quite disturbing especially if you can't. People treat you differently and try to assume what you're thinking or trying to communicate. Work out the best way to communicate. Eg use a Bluetooth keyboard with an iPad - have the iPad on view to your guest/nurse and type away all the things you're feeling and thinking.

Sleep - Sleep is a major challenge in hospital. Not only are there nebulisers and machines making noises all over the place, alarms and nurses chatting, but you're also woken for pain meds and blood pressure and so on.

Having some noise cancelling earphones is essential – eg Bose ear buds block out 80% of all sound

even without playing music, just with the power on. If someone wants to buy you a 'good luck in your operation' present this is what you want!

Eye masks are the other solution I recommend. Hospitals are never truly dark and lights are being turned on all night long. Block out the light if you can.

Finally, strike a deal with your night nurse. I decided I needed continuous sleep more than I needed my 1am pain meds and did a deal that I would press my button the moment I woke. That bought me nearly six hours of continuous sleep - absolute luxury!

Take control of your treatment. Understand what you're being given and why. Again this is about empowerment and also interesting. Means that you're part of your treatment rather than just experiencing it.

Share your experience with others who understand - namely the people in the Living with Idiopathic Subglottic Stenosis support group. The support and positive messages will be so uplifting.

Finally **be positive** – there is incredible power in positive thought - think yourself well, laugh, smile and focus on the good rather than the bad.

I'm having non-airway surgery – what should I make my surgeon aware of?

If you have to go into hospital for an operation for something other than your stenosis, then please follow this checklist:

- Advise your airway surgeon of your upcoming operation, and put him or her in touch with your other surgeon – if you can have the surgery at the same hospital your airway surgeon works at then that will be beneficial
- Talk to your surgeon about using a laryngeal mask (LMA) or an epidural rather than being intubated for your operation – avoid anything going down your trachea
- If you need to be intubated, ensure the anaesthetologist is aware they will need to use a paediatric airway tube – possibly size 4.5.
 - Insist the tube is no larger than a size 5.5 or 6, and they must use a cuff manometer to measure cuff pressure.

Preparation is essential – all doctors want to avoid causing you harm, so if you can help this situation by being your own advocate, this will be advantageous.

Pregnancy and stenosis

It is possible to get pregnant and have a family with iSGS - many patients have successfully had children with this disease.

For reasons unknown, pregnancy seems to be a trigger for some patients with iSGS. Given that the vast majority of patients affected by the disease are women, estrogen is thought to play a role in the development of iSGS. Unsurprisingly, estrogen levels surge during pregnancy. Whilst it can be scary dealing with this condition during pregnancy, the good news is it can usually be managed successfully.

Once your pregnancy is confirmed, it is important to inform the specialist treating your stenosis that you are pregnant, and also the person who will be monitoring your pregnancy about your stenosis.

It is a good idea to make sure your specialists for iSGS and pregnancy are in contact with one another. It is an added advantage if they are in the same hospital, but this is not always possible. If they are at different hospitals we recommend you make yourself aware of who will be able to help you in an emergency at both hospitals for both your stenosis and pregnancy, even if they are not currently treating you.

Unfortunately, your iSGS specialist may not have experience treating someone with iSGS during a pregnancy. To date, there are only 6-7 reported cases of management of iSGS in pregnancy in literature. Gynaecologists and Obstetricians may also never have come across anyone with iSGS, so you may need to bear this in mind when you are looking at getting advice and treatment options.

Even if you don't have any issues apart from your stenosis during pregnancy, it is advisable to arrange to deliver your baby in hospital. In case you need any kind of surgery for delivery of the baby, you are still an intubation risk and will need specialist anaesthetic care in surgery.

Airway Surgery During Pregnancy

If you need treatment on your airway during pregnancy; Don't panic! Many patients have needed steroid injections or dilation surgery throughout their pregnancy and these have usually gone smoothly with very few risks to the unborn child.



We recommend you consider a few things to ensure the pregnancy and birth goes as smoothly as possible:

- Talk to your airway surgeon about in-office steroid injections. These do not involve sedation and have proven to be a safe and harm free way to treat patients while they are pregnant.
- If in office steroid injections are not available, speak to your airway surgeon and gynaecologist/obstetrician about the optimal time to have surgery during your pregnancy. This usually somewhere between 20 and 24 weeks, but it really depends on your individual situation. Bear in mind that this may also be dictated by other factors, especially your airway and how short of breath you are.
- Make sure your specialists are in contact with each other and that your surgeon is aware of any additional information regarding your pregnancy which could impact the operation. Similarly, make sure that any medication they intend to give you during your airway surgery has been checked by your obstetrician to ensure it is as safe as possible.
- Speak to the anaesthetist department before the surgery to double check everything with them. Make sure they are aware of both your stenosis and your pregnancy and have been in contact with your specialists.
- Try to ensure an obstetrician checks the baby before and after the surgery. If you are above a certain number of weeks into your pregnancy it may be necessary have someone present during surgery to monitor the baby.

Frequently Asked Questions

What is the safest treatment for me while I am pregnant?

Airway stenosis in pregnant iSGS patients can be treated in the office with awake, unседated injections of steroid medication. This technique was reported in 2 papers in the medical literature, in 2017, and included one pregnant patient. Anecdotal evidence from some of the top airway surgeons suggests several patients have got through their pregnancy with injections alone. It may be the safest and most cost-effective way to treat the condition during pregnancy. You may ask your airway surgeon about this option and whether it is available for you.

Patients commonly receive submucosal administration of triamcinolone, dexamethasone, methylprednisolone, or solumedrol directly in the stenosis (there are no established differences between reagents). This is frequently repeated 3-5 times (at 3 week intervals) at which time patients are transitioned into clinical surveillance. Both systemic (ie. intravenous or oral) and inhaled steroids (both nasal and pulmonary) have an established track record in pregnancy (particularly in asthmatic patients), and are widely regarded as safe in pregnancy.

Do I really need surgery during pregnancy?

This is not always a simple question and is largely based on how well you are breathing and the advice of your ENT/obstetrician. Struggling through if you are really closing up and your breathing is not good could put your baby at risk. Something to bear in mind is that it is generally a lot safer

to have a carefully planned dilation than to get into any kind of emergency situation regarding breathing and/or surgery, particularly if in-office steroid injections are not available to you.

Will the anaesthetic and drugs harm my baby?

While there is obviously a risk, nowadays they have made things as safe as possible for surgery during pregnancy. Check with your doctors if you are unsure of anything. Remember that you need this surgery for a reason, and not getting enough air in could be just as harmful to the baby, if not more so. All anesthetic drugs can theoretically cross the placenta. Yet despite years of animal studies and observational studies in humans, no anesthetic drug has been shown to be clearly dangerous to the human fetus. There is no single optimal anesthetic technique.

If in office steroid injections are not available to me, can I have steroid injections after my dilation while pregnant?

It is generally felt that directly injecting them into the stenosis is the safest form of administration during pregnancy. Steroid injections may negate the need for further surgery during pregnancy.

My surgeon wants to give me a tracheotomy to get me through my pregnancy. Is this really necessary?

While this may be necessary in some cases, the majority of with iSGS during pregnancy do not require a tracheotomy during pregnancy. If your surgeon or obstetrician is wanting to place one simply because you are pregnant you may wish to seek a second opinion. The health of you and your baby is of utmost importance. If it does become necessary it may only be temporary and there will likely be good reasons why you need it.

My obstetrician wants to plan a C-section because I have iSGS - do I need a C-section or can I deliver the baby naturally?

Many iSGS patients have delivered babies naturally, so stenosis alone should not necessarily be a reason why you can't have a natural birth if you choose to. That being said, there may be other additional reasons why a C-section is recommended, and it is important to follow the advice of your doctors on this.

Will I need to be intubated to deliver my baby?

The good news is that most planned surgery for delivery is usually done using other methods such as an epidural, which won't require intubation. However, it is a good idea to get a note from your ENT about what is needed in terms of anaesthesia, specifically, what size endotracheal tube is needed if you must be intubated in the event you need any emergency surgery.

Tips for dealing with iSGS during pregnancy

If you are taking any medication for your stenosis make sure that you check with your doctor if it is safe to use during pregnancy. If you have a lot of mucus, nebulising with cool boiled water or saline might help as an alternative to expectorant medication. See our section on reducing mucus naturally.

Make sure you carry a note with brief details of your stenosis and pregnancy, and what to do with your airway in the event of an emergency. Usually, this contains suggestion regarding avoiding intubation, use of a small size breathing tube (4.5-5.5 usually) if intubation is necessary. Remember to include details of your doctors and how to contact them.

If you think you feel out of breath, don't wait to see your doctor. Some patients close quite quickly during pregnancy and you may want to get it checked out.

A few weeks before your due date, have a check-up with your airway doctor so you have an idea of how your airway is looking and how open or closed you are before you deliver. Use of a peak flow meter during your pregnancy may be helpful to track the size of your airway.

Try to relax! While it can be stressful and scary dealing with this condition during pregnancy - try not to get worried about it.

Research in progress

A major study into understanding the treatments and efficacy of treatments for iSGS is being carried out by Vanderbilt University in Nashville Tennessee. iSGS patients from around the world are taking part in this research and I am a patient partner in this process.

Can I take part in this research?

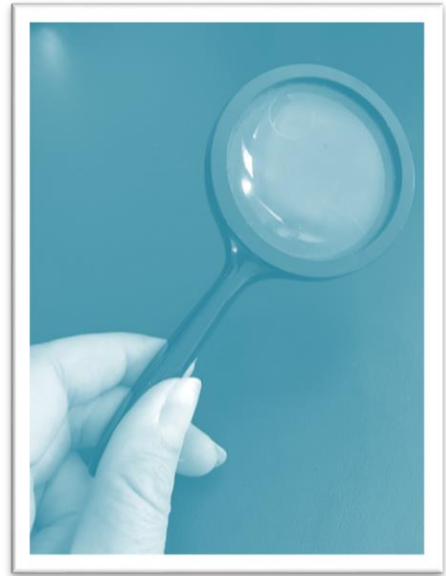
Admission to this research has now closed. There are more studies happening all the time – if you are willing to take part in upcoming research regarding iSGS, share your email address with Catherine via idiopathic.subglottic.stenosis@gmail.com

Latest findings

Recent research by Vanderbilt University (June 2016) has shown that iSGS patients have very high levels of inflammatory cytokine IL-17 in their airways. Further investigation to understand what might be driving this, has found that iSGS patients have Mycobacteria present. Doctors are currently unsure of the complete identity of this bacteria, but can safely say it is NOT Tuberculosis, although it comes from the same very large family (a distant cousin perhaps).

At this stage, doctors are unsure which came first – the scarring or the bacteria, but this is their next stage of research.

The next update of the Vanderbilt study is due in September 2018.



Appendix

North American Airway Collaborative (NoAAC) PR-02 ISGS trial funded through PCORI

In May 2015, we heard the great news that the North American Airway Collaborative was successful in its quest for funding from the USA government for research into idiopathic subglottic stenosis.

The collaborative is made up of doctors from predominantly across the USA but also including the UK and Australia. The founder of the Living with Idiopathic Subglottic Stenosis group, Catherine Anderson, was also recruited into the collaborative as a consultant to assist with research and to coordinate research among patients with iSGS who are not participating in the study with their surgeon.

FREQUENTLY ASKED QUESTIONS

What is the aim of the trial?

The trial aims to:

- Refine doctors' epidemiology on iSGS patients – ie helps them to understand any patterns, causes, and effects on health
- Establish baseline quality of life metrics for the disease (4 electronic measures in voice/swallowing/breathing/ and quality of life)
- Understand the psychological effects of having iSGS – ie understand more about what social support is needed, communication with providers, and disease fears
- Measure the time to recurrence after receiving a procedure
- Investigate the changes in quality of life that accompany treatment

Which doctors are taking part in the research?

See the document under the Files tab in the group: North American Airway Collaborative - List of Participating Centres - 10-05-15.pdf which has the latest list of participants. There are also some additional centres which are applying to take part who are not yet on the list – including Dr David Veivers in Sydney Australia.

My doctor/centre is not listed there – can I still take part?

The study is now closed to new participants.

What was involved?

Frequent questionnaires and follow up questions after a surgery to understand how you are feeling. You will be asked to allow your doctor to share your medical records plus any test results with the team.

Who is taking part?

The trial was open to all patients with idiopathic subglottic stenosis.

I have no or limited insurance. Will the NoAAC help pay for my treatment?

Unfortunately not – this study is a research project and not a charitable foundation for patients. Everyone receives the 'standard of care' they would typically obtain with their provider. There are no additional tests or procedures involved in the PCORI trial, thus the trial has no additional associated costs to you.