Living with iSGS
Idiopathic subglottic stenosis

IDIOPATHIC SUBGLOTTIC STENOSOIS
A ROUGH GUIDE
FOR BEGINNERS

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Last updated: October 2022

Support: Facebook.com/groups/AirwayStenosis | **Twitter:** @SparkySparkler

Foreword – Welcome to the ‘Rough Guide’

Welcome to the ‘Rough Guide’ – this is designed to be a patient’s introduction to subglottic stenosis, aiding your understanding of this diagnosis, busting some myths, and helping you to take control of your health and treatment. We share tips to empower you in your journey; listing the recommended tests your doctor should be doing, questions to ask your doctor in different situations, advice, equipment, advice that will make living with this diagnosis easier, and simple ways to monitor your breathing.

I am Catherine Anderson, and I was diagnosed with idiopathic subglottic stenosis in 2004 after two years of misdiagnosis. I know the relief you feel when you find there IS something wrong with you, followed by the confusion when you try to understand this prognosis.

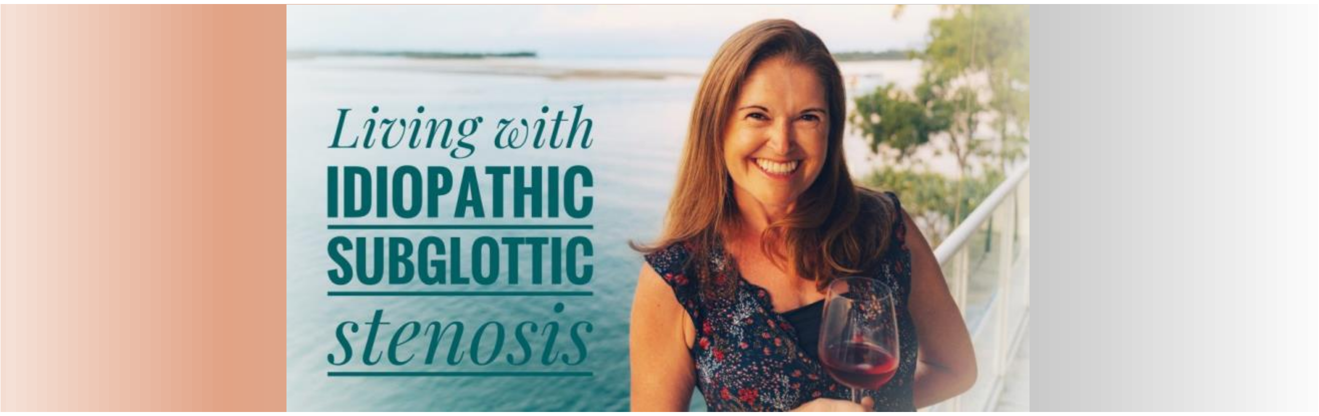
In 2009 I founded a support community ‘Living with idiopathic subglottic stenosis.’ Learning from negative experiences with other groups, this one encourages members to collectively learn and keep the discussion fresh and solution focused, avoiding repetitive questions or revisiting things we know the answers to. The community is a big contributor to research by Doctors around the world, enhancing knowledge about our disease. This guide is designed to help accelerate new patients’ knowledge so they can ask more informed questions as well as join in with research, empowering them to be a part of the solution.

This disease is so rare that many doctors, even those specialising in ear, nose and throat conditions, often have little experience with it, meaning we patients **have to self-advocate** to ensure we get the right treatment. Everything in this guide has been approved by the most experienced doctors treating this disease, so you are reading and sharing the most accurate information.

I know this diagnosis can be frightening. Hopefully, this document can ease your fears and help you to live as normal a life as possible, being better informed and working in partnership with your doctor to get the best treatment for you.

Wishing you easy breathing!

Catherine



Executive summary – the quick guide

While this guide is packed with useful information about subglottic stenosis, we appreciate it can sometimes be too much to read all at once, and details can be missed. Here is the top-line summary of what you should know right now.

What is subglottic stenosis (SGS)?

A **narrowing of your airway caused by the formation of scar tissue just below your vocal cords**. It is rare, with just two people in a million diagnosed each year, though since Covid-19, numbers have been increasing among those treated with intubation.

Why have I got this?

There are a few potential causes, including damage from a **prior intubation** (e.g. during a previous operation), damage from **inhaling chemicals or heat**, an **autoimmune disease** (such as GPA or lupus) or from an **unknown cause**. Idiopathic literally means 'cause unknown'. Your doctor should investigate all avenues as the treatment will slightly differ if a cause is detected.

Is it life threatening?

Yes, this **must be taken seriously**. Your biggest risk is a plug of mucus blocking your airway while narrow. Share information with family and work colleagues that can help save your life if your airway should block.

It's important that you **talk to your treating doctor** about **flagging your record** to **prioritise your appointments** should you need treatment in an emergency. This disease can be unpredictable, and you do not want to wait weeks for an appointment if you are struggling to breathe. Receptionists and schedulers do not always realise the risks associated with SGS.

How is it treated?

Initially, it is likely you will have a **dilation surgery**. This is usually day surgery where the doctor operates through your mouth to open up the scar. You will wake up breathing well, either right away or within a day or so. You may have a sore throat, and maybe your chest and shoulders will feel a little stiff for a couple of days. Usually patients go home the same day and can be back at work in 2-3 days. **Other treatments** include **steroid injections** (done in your doctor's office) or **more major surgeries such as resection or the Maddern procedure**. Please be certain you are comfortable with your doctor's experience and expertise before following a treatment path. You will need to self-advocate for your airway health.

What should I do to help my airway?

A **nebuliser** (a machine aimed at delivering moist air to your airway) is essential – you can use it with saline to moisten your airway and help move mucus and prevent mucus plugs. A silent, portable one is what most people prefer to use. You can buy these online.

A **peak flow meter** is a cheap and easy way to monitor your breathing at home, without the need for a doctor's appointment and that uncomfortable scope (camera looking at your airway via your nose). There are several apps to help record your readings, and this can help you learn about your airway – what it is like when you are breathing well, and when it is best to contact your doctor.

Keep **hydrated** and eat a **healthy diet**, minimise sugar and processed foods. Ideally follow an anti-inflammatory diet. Within reason, **keep yourself fit** and **maintain a healthy weight** – all will help you cope better with the symptoms of this rare disease.

It is recommended you are **immunized** for flu, pneumococcal and Covid-19. This will help prevent additional complications which could prove life threatening when coupled with subglottic stenosis.

What is idiopathic subglottic stenosis?

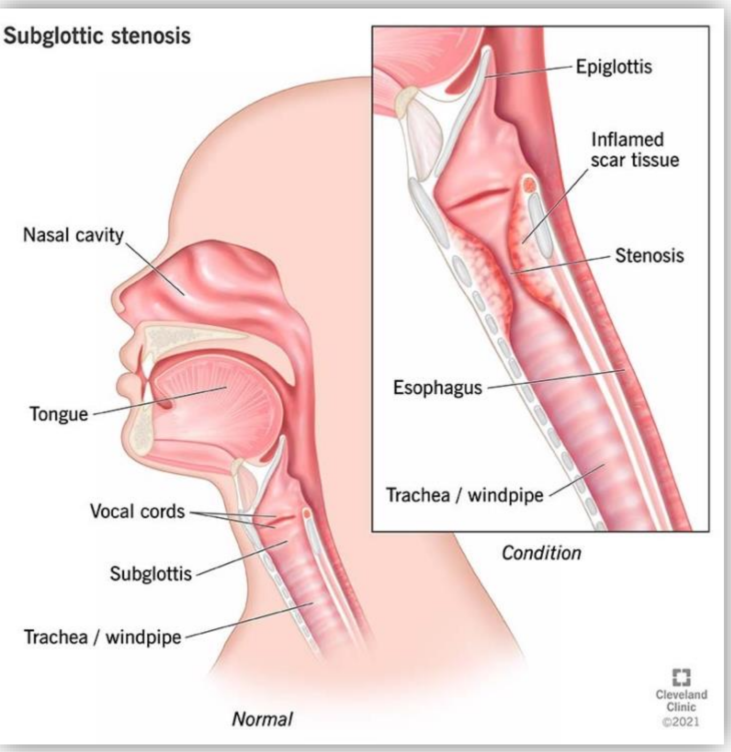
So, you have just been diagnosed and it's all very overwhelming. Suddenly doctors are reeling off words you, your friends and family have never heard of, and when you search on the internet there is no or very little insight to be found. Well done for finding this document and hopefully our Facebook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

Getting down to basics;

- Idiopathic – is a fancy way of saying 'no known cause'
- Subglottic – is the part of the larynx below the vocal cords (glottis). It connects to your trachea ('windpipe'). 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. The other types of stenosis may result in different treatment paths.



This document will help you regardless of your stenosis location, and of course, even if your stenosis is not idiopathic, you will still learn a lot here.

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 a referral to another doctor**. Your airway is not to be experimented with. Unfortunately, not all ENT and Thoracic doctors are experts or fully familiar with the best way to treat subglottic stenosis.

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 (see below – iatrogenic stenosis)
- Thyroid or major anterior neck surgery
- Exposure to radiation on your neck
- Caustic (e.g. 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)

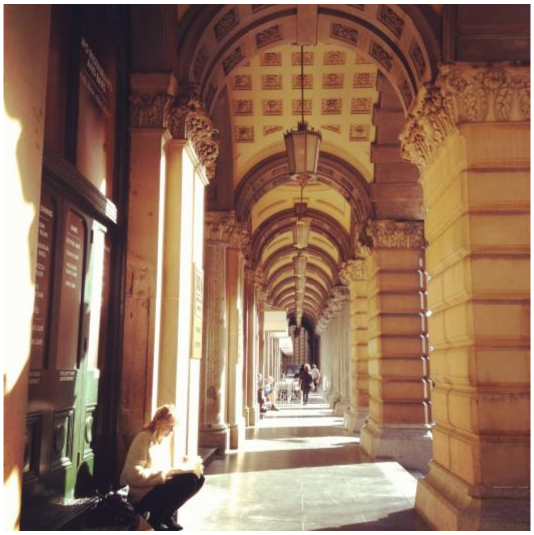
Autoimmune - Patients with documented diagnosis of Wegener's (GPA), Relapsing Polychondritis (RPC), Systemic Lupus Erythematous (SLE), Rheumatoid Arthritis (RA), Epidermolysis Bullosa (EB), Sarcoidosis, Amyloidosis or Mucous Membrane pemphigoid (MMP). Doctors may also treat you as autoimmune if your blood tests positive for ANCAs (Antineutrophil cytoplasmic antibodies) or if your stenosis 'behaves' like an autoimmune stenosis (high levels of inflammation and/or returning rapidly post surgery)

Polytrauma - Patients 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 (intubation in intensive care) or a tracheostomy – either immediately or within 2 years of intubation (although if you have EVER been intubated, it is certainly worth getting hold of your hospital notes to see whether there were any difficulties – there is a chance your stenosis was caused by the intubation).

What are the common symptoms of SGS?

We all share some common symptoms:



Stridor – this is what we often describe as a wheeze, the sound when we breathe in and out. 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 are 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 – we generally do not have any more mucus than the average person; rather it becomes harder to shift because of the scar tissue and the narrowing (see diagram). If you have inflammation in your airway, it is likely there is more mucus as the two (inflammation and mucus) come hand in hand. 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 are concerned, see a doctor, but mostly this clears up. Please see our **tips for thinning and minimizing** mucus and the **section on ‘mucus plugs’**.

A narrowed airway requires more rapid airflow to maintain oxygenation. The mucous layer evaporates leaving a dry sticky residue and low mucosal temperature stimulates nerves to cause cough.

Inflammation – all this coughing and shifting of mucus can lead to inflammation. If it gets 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), investigate 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. Chronic inflammation can be a sign of autoimmune disease – ensure you are tested annually for the suite of blood-tests detailed later in this document.

Fatigue – as it becomes more challenging to breathe, you are likely to feel more tired. You will feel that your 'get up and go' has 'got up and gone'! Everything you do is likely to feel more of a challenge and you may find yourself feeling tired during the day. This should ease after a dilation – in fact, many patients report temporary feelings of euphoria once they can breathe well.

Inability to concentrate – alongside fatigue, the more you are concentrating on breathing, the less space you will find in your head to concentrate on other things in life – essentially you are in survival mode. After your airway has been opened back up, you should find space in your head for things other than breathing!

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**!

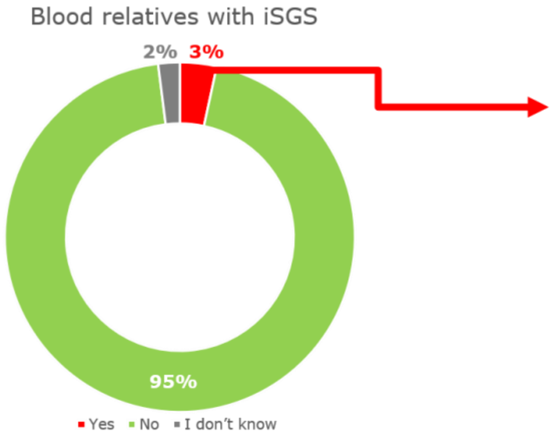
Who gets SGS?

Idiopathic subglottic stenosis is exceedingly rare – less than one in 400,000 people get this disease (the definition of 'rare') – it is in fact estimated to only impact one person in half a million per year.

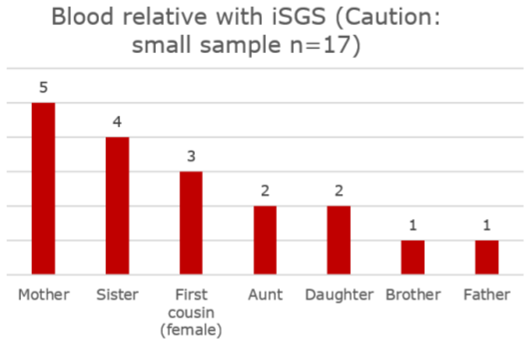
If you are female, then you are one of many, as this affects 98% females. If you are male, then you are truly special and part of the 2%.

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. The big message doctors need to get is that with asthma, the wheezing sound is heard when breathing out, with a tracheal/upper airway blockage (rather than a lung) issue, the wheezing sound is also heard when breathing in.

People are often afraid of passing this on to their children – that is very unlikely. Just 3% of SGS patients have a blood relative with this disease, suggesting this is not something you should be too worried about. If a relative (particularly a female) has similar symptoms to you, then they should suggest this as a diagnosis to their doctor and ensure they get sent to an ENT for investigation.



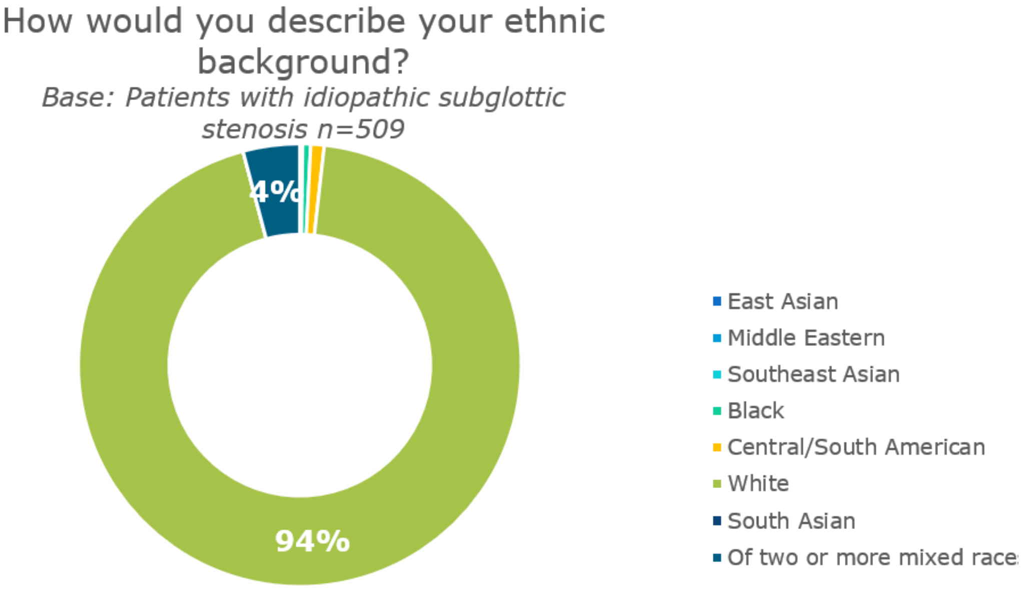
Q30. To your knowledge, have any other blood related family members been diagnosed with a stenosis (eg brothers, sisters, parents, aunts or uncles who are directly related to your parents, grandparents)? Base: Patients with idiopathic subglottic stenosis, n=502



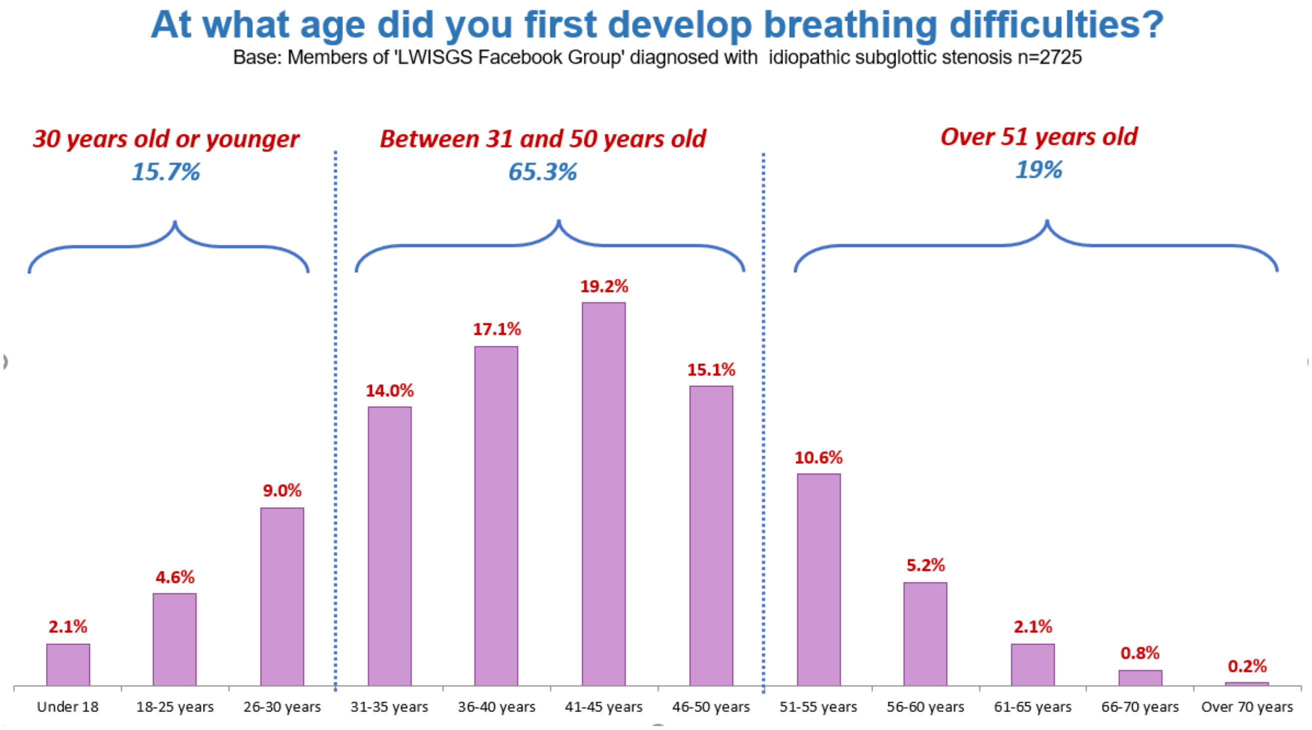
Q31. What type of subglottic stenosis were they diagnosed with? Base: Patients with idiopathic subglottic stenosis who indicated they had a blood relative also with a stenosis n=17

Ethnicity and heritage

More than nine in ten patients describe their ethnic background as white, suggesting there may be a genetic element to this disease:

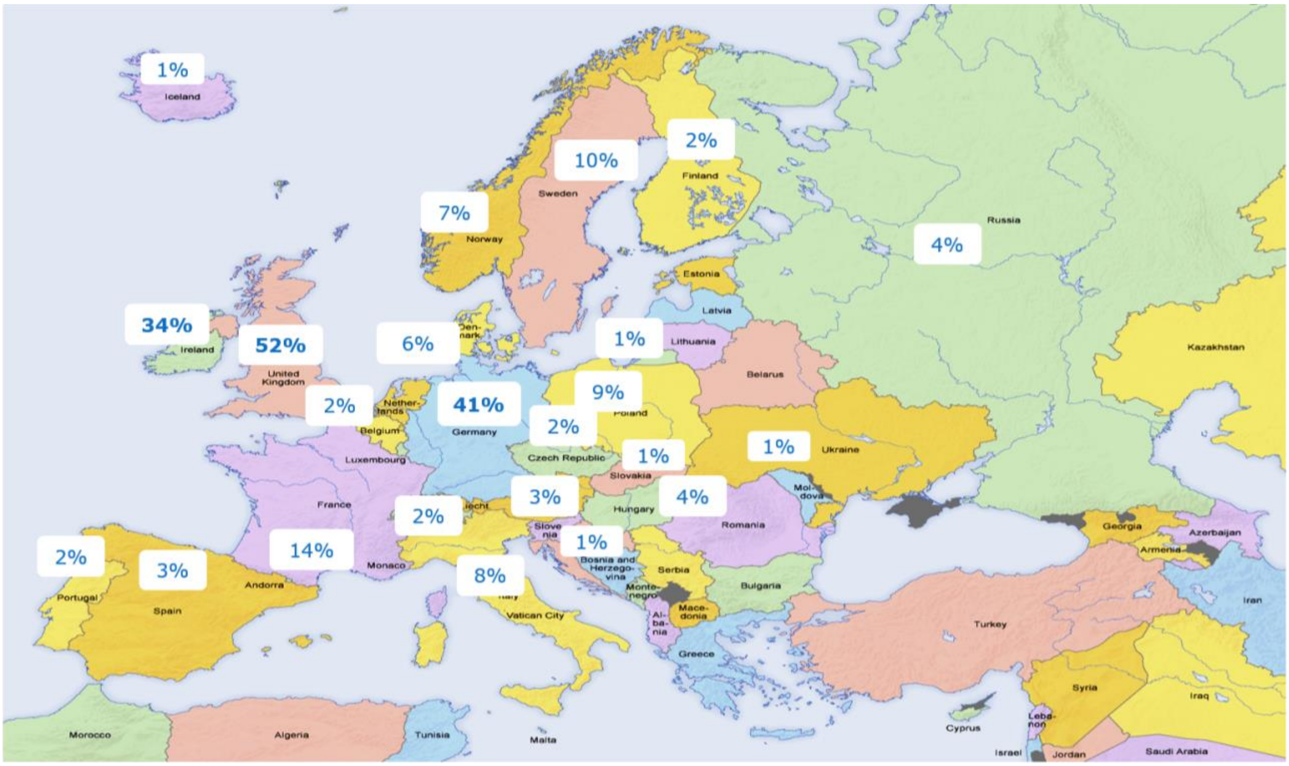


One in two patients experience their first symptoms between 31 and 45 years of age.



Patients with SGS are no different from the rest of the population in terms of other health conditions (heart, thyroid, cholesterol etc), and other than breathing (!) are healthy.

More than nine in ten have a European ancestry:



Q11. Please explain more about your European heritage – from which country or countries?
Patients with idiopathic subglottic stenosis with ancestry in Europe n=425

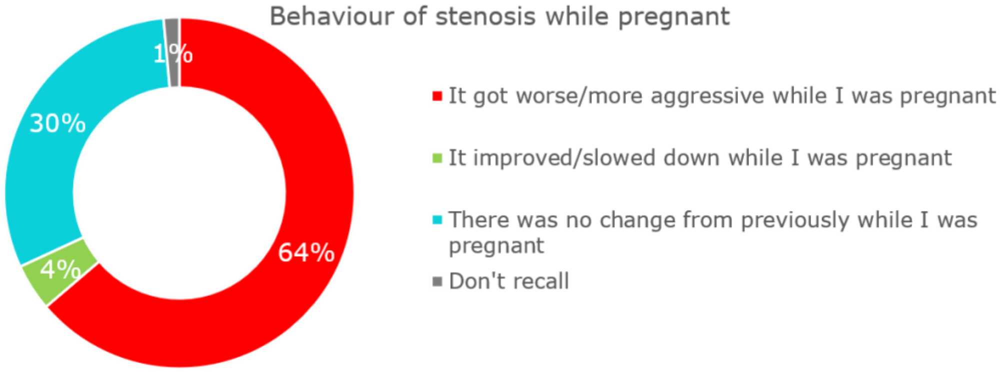
Hormones

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

17% of women with SGS have never been pregnant, and only one in five (22%) has ever been pregnant while they had SGS. For 26% of women, their stenosis did not appear until after menopause.

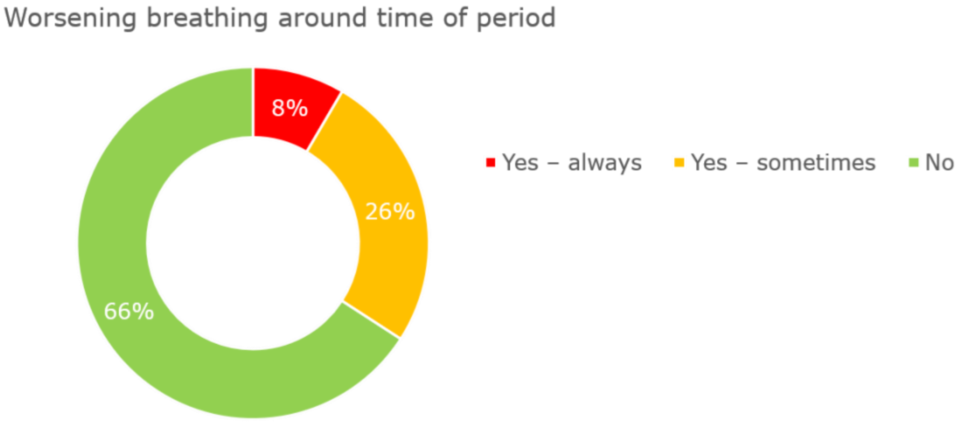
The stenosis first appeared for around one in three (30%) while they were pregnant.

For those women who **already have a stenosis when they get pregnant**, three in five (64%) see the stenosis get worse during their pregnancy. One in three (30%) see no change.



Q51. How did your stenosis behave during your pregnancy? Base: Patients who got pregnant when already diagnosed with idiopathic subglottic stenosis, n=69

Looking only at women who have not reached menopause, around one in three (34%) finds their breathing gets worse around the time of their period. This may be a result of increased CRP levels around the time of women’s periods*.



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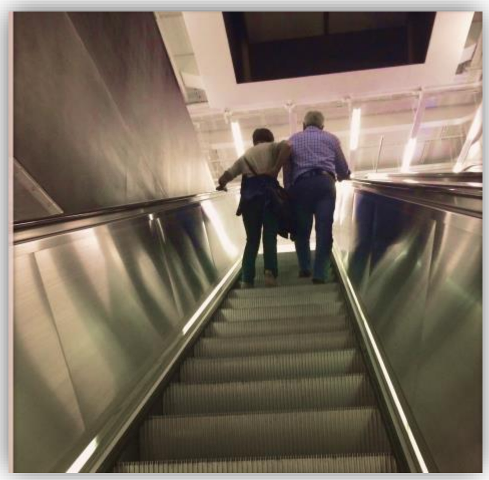
*Journal of Women's Health VOL. 25, NO. 9 | The Association of Inflammation with Premenstrual Symptoms - Ellen B. Gold, Craig Wells, Marianne O'Neill Rasor - Published Online: 1 Sep 2016 https://doi.org/10.1089/jwh.2015.5529

Debunking the myths about SGS

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



"SGS is caused by reflux" – No. Your SGS is NOT caused by reflux. Think about it. About 60% of the world's population has reflux – males as much as females – and yet SGS 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 being referred for further evaluation. A variety of diagnostic tests are available (e.g. esophageal pH probe testing, esophageal impedance testing, esophageal manometry, pharyngeal probe testing – called ResTech, pharyngeal pepsin assay, esophageal endoscopy, etc), depending on where you live. 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 do not need any more health problems, do we?!). There is evidence acid reflux may slow or prevent adequate healing after endoscopic or open procedures, so it is important to diagnose and treat reflux in this situation.

"People with SGS have more mucus than 'normal people'" – the fact is we generally have 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 must 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 or steroid injections** to avoid the potential of a plug you cannot shift. If you have chronic inflammation present you *may* also have more mucus than normal – reducing inflammation (eg through diet) should also help reduce excess mucus.

"I cannot exercise because I have SGS" Many of us make ourselves continue to exercise despite SGS. 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, swim, or go to the gym several times a week despite a narrow 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 SGS.

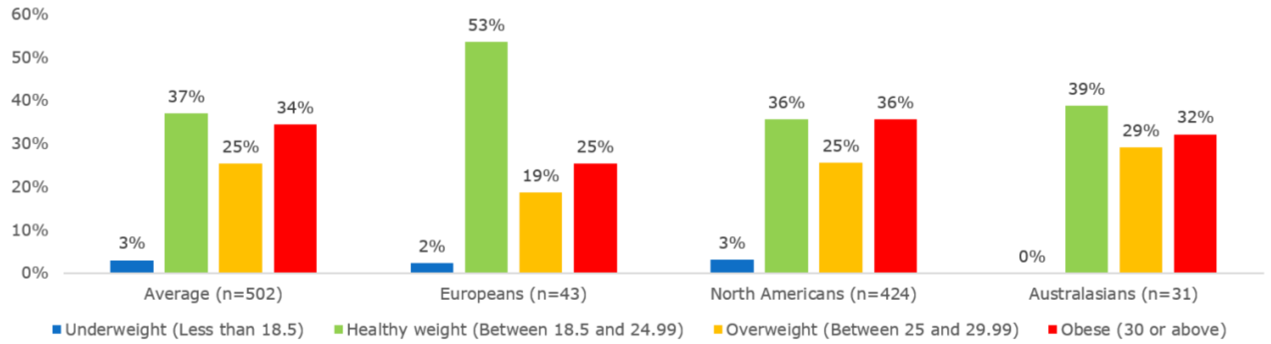
We suggest you start exercising as soon as you are breathing well after a dilation and keep going as long as you can. Just do what you can within your limits and any other conditions you have.

"I am overweight because of SGS" – 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 SGS is not helping. Work hard to reduce weight naturally – cut back on sugar and processed foods and replace with fresh vegetables.

Body Mass Index (BMI) helps us to understand the general health of people by looking at their weight versus their age, gender, and height. This is important to look at because obesity has a negative impact on health overall*. Obesity is associated with hypertension, dyslipidaemia, ischaemic heart disease, diabetes mellitus, osteoarthritis, liver disease, and asthma. Obstructive sleep apnoea (OSA) is a common problem in the morbidly obese.

When coupled with a chronic disease such as idiopathic subglottic stenosis, risks are increased further. Every time an obese patient goes under an anaesthetic, they are at increased risk of airway collapse – especially if they have had symptoms of sleep apnoea or snoring.

One in three (34%) SGS patients is obese.



Q28. What is your Body Mass Index (BMI)? (Calculator provided) Base: Patients with idiopathic subglottic stenosis, n=502

**National Institutes of Health. National Heart, Lung, and Blood Institute. Clinical guidelines on the identification, evaluation, and treatment of overweight and obesity in adults—The evidence report. Obes Res 6(Suppl 2):51S–209S. 1998. Anaesthesia and morbid obesity - Sharmeen Lotia, MBBS MRCP FRCA Mark C. Bellamy, MBBS MA FRCA Continuing Education in Anaesthesia Critical Care & Pain, Volume 8, Issue 5, 1 October 2008, Pages 151–156

Recent research[1] has also found that SGS patients with obesity (particularly with a BMI of between 30.0 and 34.9) are likely to see their stenosis reoccur much faster after a dilation than those patients of healthy weight or underweight. Another good reason to consider reducing your weight.

If you need to calculate your BMI you can use this calculator – you will need your weight, height, age, gender, and an idea of your activity levels - https://www.nhs.uk/live-well/healthy-weight/bmi-calculator/

"SGS is caused by being pregnant or taking hormones" – this cannot be totally debunked but given that around one in five patients with SGS has never been pregnant, that suggests it is not the only reason. One in ten SGS patients has never taken any hormones (e.g. the pill). We also have males diagnosed with SGS who have not been on the pill or pregnant.

"You must not drink alcohol or caffeine with SGS" – in moderation, alcohol and caffeine are fine. We recommend you balance any drinking of alcohol or caffeine with plenty of water – dehydration is a real problem with SGS and can lead to life threatening complications such as mucus plugs. Of course, if you have other medicines or health conditions that require you to avoid these substances, then always listen to your doctor's advice, otherwise, hydration is key.

"Having major surgery (e.g. resection, Maddern) will cure me of SGS" – Unfortunately, there is no cure to SGS at present. At best, major surgery may put your symptoms into remission, possibly for 10 or more years. You will always live with the diagnosis of subglottic stenosis, and there is a chance it will come back. Your likelihood of achieving a longer time without regrowth is directly correlated to the experience of the medical centre and team performing your surgery – the more experienced the centre, the longer your airway is likely to be stenosis free.

"I don't need to have blood tests for ANCA, ANA etc because I tested negative in the past" – doctors recommend you have these tests annually even if you have tested negative – ideally when you have noticed your scar growing back or around the time of a dilation. The result can change even after many years. You can usually organise blood tests from your GP/primary health care practitioner.

"Your oxygen levels will be low with subglottic stenosis" – while a difficulty breathing might suggest oxygen would be low, for most airway stenosis patients oxygen is not an issue. Our bodies are simply amazing, becoming very efficient at extracting the right amount of oxygen for survival, despite the narrow airway. If you are experiencing low O_2 levels, there may be another health condition at play that needs investigating.

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[1] Evaluating the Association of Clinical Factors With Symptomatic Recurrence of Idiopathic Subglottic Stenosis May 2019 JAMA Otolaryngology - Head and Neck Surgery 145(6) DOI: 10.1001/jamaoto.2019.0707

What tests should my doctor be doing?

There are a range of blood tests that should be regularly run to check for potential autoimmune causes of your subglottic stenosis.

There is not a definitive list of tests, but 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.

- **ESR (Erythrocyte sedimentation rate)** - used to detect and monitor the activity of inflammation as an aid in the diagnosis of the underlying cause.
- **CRP (C-reactive protein)** - used to identify the presence of inflammation, to determine its severity, and to monitor response to treatment. A more sensitive form of the test, high-sensitivity C-reactive protein (hs-CRP), is used to assess your risk of heart disease.
- **RF (Rheumatoid factor)** - to help diagnose rheumatoid arthritis (RA) and Sjögren's syndrome
- **ANA (Antinuclear antibody; fluorescent antinuclear antibody)** - screen for certain autoimmune disorders, such as systemic lupus erythematosus (SLE), polymyositis, and a number of others
- **ACE (Angiotensin converting enzyme)** – screens for sarcoidosis.
- **ANCA** - looks for antineutrophil cytoplasmic antibodies (ANCAs) in your blood test for vasculitis related autoimmune diseases, such as Granulomatosis with polyangiitis (once called Wegener granulomatosis), Microscopic polyangiitis and Eosinophilic granulomatosis with polyangiitis (once called Churg-Strauss syndrome).

Your doctor should also take a biopsy of the tissue while under anaesthetic which will be tested for signs of granulomas (small areas of inflammation). Granulomas form when the immune system attempts to wall off substances it perceives as foreign but is unable to eliminate.

It is strongly recommended that the 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.

If your surgeon suspects laryngeal reflux (acid from your stomach reaching your airway) then ask your doctor to refer you for further investigation. A variety of diagnostic tests are available (e.g. esophageal pH probe testing, esophageal impedance testing, esophageal manometry, pharyngeal probe testing – called ResTech, pharyngeal pepsin assay, esophageal endoscopy, etc), depending on where you live. In the event you test positive, taking 20ml of liquid containing sodium alginate daily has minimal side effects. (In the UK, Australia, and USA, use Gaviscon reflux medicine, available at most pharmacies)

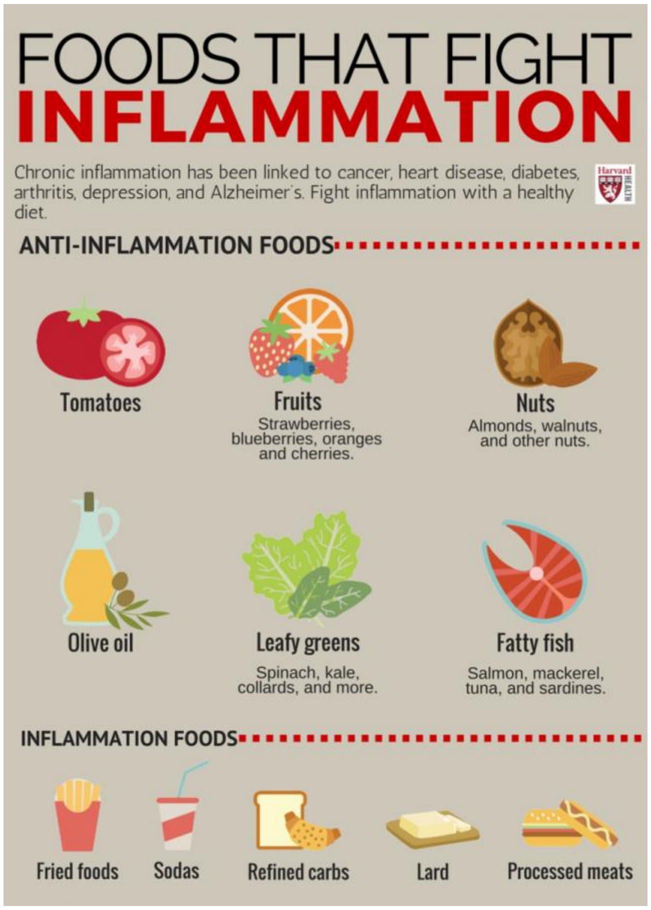
Tips to help reduce inflammation, mucus, and coughing

Coughing is a fact of life with SGS. Your airway has scar tissue instead of healthy mucosa. In your airway, the mucus gets stuck in the region of the scar tissue, meaning it becomes thick and can potentially harbour bacterial infections and build up into a mucus plug (see next section). Inflammation is also often an issue for airway stenosis patients which can exacerbate the issue. Fortunately, there are some things you can immediately do to help with this.

Diet

What you eat has a massive impact on your breathing – both the inflammation and mucus elements. You can search on the internet for the most current advice, but generally:

- Eat a diet rich in fruits and vegetables, especially:
  - Green leafy vegetables – such as kale, spinach, cabbage, water cress, romaine lettuce, Swiss chard, arugula, and endive
  - Dark yellow vegetables – such as pumpkin, yellow peppers, beans, and carrots
  - Whole nuts -
  - Fruits – especially blueberries, pomegranate, orange, cherries, strawberries, apples, and pears
- Drink– tea, coffee, and red wine (in moderation), and water (as much as you like)
- Cook with and dress salads with extra virgin olive oil.
- Fatty fish – e.g. rich in omega 3 fats in sardines, mackerel, salmon
- Nuts – especially walnuts.
- Apple cider vinegar – one tablespoon in soda water daily



FOODS THAT FIGHT **INFLAMMATION**

Chronic inflammation has been linked to cancer, heart disease, diabetes, arthritis, depression, and Alzheimer's. Fight inflammation with a healthy diet.

Minimise

- Red meat, processed meat, organ meat (kidney, heart, lungs etc) and refined carbohydrates (white flour and white rice)
- Sweetened beverages (anything with sugar in particular)
- Dairy products and soy – many members notice an immediate difference when cutting out cow's milk and its associated products (i.e. cream, yoghurt, butter, cheese, and products containing milk derivatives such as milk protein, lactose, whey). Soy milk is often portrayed as an alternative to cow's milk but can often result in similar mucus effects.

Other solutions

- Nebuliser - with 0.9% normal saline or cool boiled water - 5ml at a time as often as you need it. Research has found that 7 in 10 SGS patients experience improved mucus symptoms, and 6 in 10 experience improvement in cough (K Tanner, 2019).
- Humidifier in heated or air-conditioned environments

Over the counter medicines

- Mucinex or Robitussin Chesty Cough Capsule or liquid. Vicks cough syrup
- Bisolvon Chesty tablets (Australian brand) - contains bromhexine hydrochloride 8mg (breaks down mucus to make it easier to clear)
- Difflam (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.
- Nasacort (brand name in Australia, USA & UK) or Telnase 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) - do not use long term (due to negative steroid side effects) or when you have a throat infection.
- Antihistamine can help dry existing mucus and prevent additional formation due to allergy.

Prescription medication

- Prednisolone / oral steroids

Techniques to help clear mucus

Deep Coughing: Start by taking a deep breath. Hold the breath for 2-3 seconds. Use your stomach muscles to forcefully expel the air. Avoid a hacking cough or merely clearing the throat. A deep cough is less tiring and more effective in clearing mucus.

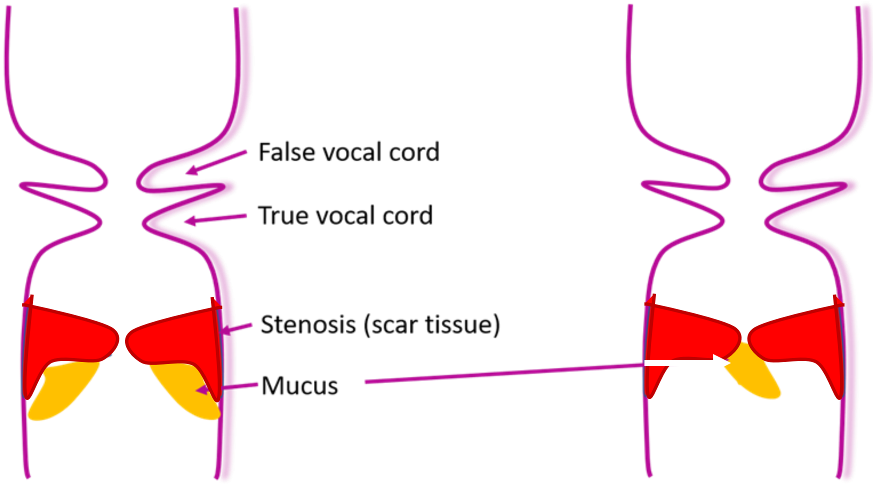
Huff Coughing: Huff coughing, or huffing, is an alternative to deep coughing if you have trouble clearing your mucus. Take a slow deep breath that is slightly deeper than normal. Use your stomach muscles to make a series of three rapid exhalations with the airway open, making a "ha, ha, ha" sound. Follow this by controlled diaphragmatic breathing and a deep cough if you feel mucus moving.

Nebulizer Use in Adults with Subglottic Stenosis: A Survey Study, Ann Otol Rhinol Laryngol 2019 Apr;128 (4):345-351. Doi:10.1177/0003489418823797. Epub 2019 Jan 14. K Tanner, C Anderson, M Smith

What is a mucus plug and how do I avoid it?

A mucus plug is a potentially life-threatening incident for people with subglottic stenosis and **must be taken very seriously**.

What is a mucus plug?



If you have never experienced a mucus plug, the following diagram attempts to explain what is happening.

Your stenosis is creating an unnatural obstruction in your airway. Even if you have a dilation, the lining of the airway still does not function properly at the site of scar. Scarring damages the delicate cilia that help whisk mucus out of the airway. This can lead to mucus getting stuck in your airway. You will have to cough to make this shift, and ideally cough it out (or swallow it).

Sometimes though, this mucus can get more difficult to cough out. This can happen when you get dehydrated, or when you get sick. If you have an infection you might find that instead of white or clear mucus being coughed up, it is yellow, green, or orange. This is a sign you are at risk of a mucus plug forming. When infected with bacteria, it can become quite thick, sticky, and hard, and is particularly challenging to cough.

When it becomes big enough, there is a risk of this blocking (plugging) your narrow airway. This is quite terrifying when it happens, and usually your survival instinct kicks in and you can manage to somehow cough it out.

Should a mucus plug affect you, you will not be able to speak and tell others what is happening, so it is best to forewarn those around you that this is a risk.

What if I cannot shift the plug?

If you cannot shift the plug, you will not be able to breathe or speak and will fall unconscious. If you have somehow managed to alert someone that you are struggling, they **must immediately** call for emergency medical assistance. Speed is of the essence here, and again it is important people around you are prepared with what to tell medical professionals when they call.

- You are not breathing and are now unconscious.
- Your airway is blocked.

At this stage, the name of your disease is **not** the most important thing, getting air is primary. Make sure they know what number to dial (e.g. 911/000/999 depending on the country they are in) – it is amazing how many people have a mental block to what the correct emergency services number is when they need to call it.

An option those around you can try is the **abdominal thrust** (also known as the ‘Heimlich manoeuvre’). This is a first aid technique used to move foreign bodies caught in your airway in an emergency. This may dislodge the plug and can be tried while waiting for emergency services.

If you are alone, you can also attempt this on oneself:

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Mouth to mouth resuscitation can also be attempted by those around you, they can potentially push the plug back down into the airway allowing you to breathe. This can also be tried before you lose consciousness.

1. With the airway open (using the head-tilt, chin-lift manoeuvre), pinch the nostrils shut for mouth-to-mouth breathing and cover the person's mouth with yours, making a seal.
2. Give two **strong** rescue breaths...
3. Proceed with chest compressions to restore circulation.

How do I prevent a plug from happening?

Vigilance is key to prevention. If you notice the early signs, you need to act. These could be one or more of the following:

- Coughing more than usual
- Coughing up small lumps of mucus
- Mucus, which is yellow, green, or orange
- Your peak flow dropping 40%+ suddenly when it has usually been stable.

If you notice any of these, your first step should be to use your saline nebuliser **at least** twice a day. Adding moisture to your airway can help you cough up the growing plug before it becomes unmanageable. Staying hydrated with enough water is also extremely helpful.

In addition, if you have noticed green, yellow, or orange coloured mucus, then consult with a doctor about using antibiotics to help clear any infection.

Finally, keep monitoring your breathing with your peak flow meter – if it remains low despite action, then contact your ENT surgeon, and if you continue to have concerns, go to emergency at your local hospital.

A mucus plug is very serious, and subglottic stenosis patients have died from this.

Should I be worried about my mucus/phlegm?

Not all mucus/phlegm is of concern, so here is a guide to help you understand whether you are potentially at risk from a mucus plug.

Clear: good news, this is healthy mucus. Keep yourself hydrated and nebulise with saline as necessary (every two to three days or more often if you need). Low risk of creating a mucus plug.

White: a virus such as the common cold. Increase your hydration, avoid caffeine and alcohol, and use a saline nebuliser regularly (daily or more often if you need). Not usually a risk for plugging but keep an eye on things – if it thickens or becomes hard to cough up, then increase your nebulising frequency.

Yellow or green: a bacterial infection. Yellow phlegm suggests that immune cells are starting to work at the site of the infection or another type of inflammatory condition. White blood cells are the cells of the immune system that are responsible for fighting germs. As they continue to fight the infection, the phlegm picks them up, giving it a yellowish tinge.

If accompanied by a fever, then see your doctor for antibiotic treatment.

Increase your hydration, avoid caffeine and alcohol, and use a saline nebuliser regularly (daily or more often if you need). Can be a risk for plugging, so vigilance needed to keep it moving with fluids.

Orange or brown: sign of inflammation and dried blood. Increase your hydration and nebulise more frequently with saline. See a doctor if you are coughing up lots of bright red blood, but small smears are not of concern. There are many reasons for blood in the phlegm. A lot of coughing, such as with subglottic stenosis, can sometimes cause small blood vessels in the lungs or airways to break and bleed.

When there is swelling in a person's nasal passage, they can get a nosebleed. This can cause blood to seep into postnasal drip that they then cough out. Can be a risk for plugging, so vigilance needed to keep it moving with fluids.

Texture: Thicker/tackier mucous is of highest risk to airway stenosis patients. This is most likely to increase your risk of a plug. Reducing your risk is about:

- Nebulizing saline – delivers moisture directly to your airway
- Running a humidifier - can help moisten the air you breathe

Both options make it easier to cough and loosen up the phlegm stuck in the airway.

Some individuals may find that gentle walking can help them cough up excess phlegm.

Your essential medical emergency kit

The next section details what **equipment will help daily**, but we also recommend you have an **emergency medical kit** (or prescriptions you can fill quickly) to hand so you can act quickly when required. This is particularly important if you are travelling and don't have access to your usual doctor(s). It is always a challenge to explain this disease to someone who is not an expert, so being self-sufficient is important.

Speed is often of the essence when it comes to keeping you and your breathing safe, and you do not want to have to wait for appointments or calls to be returned.

Vaccinations

Three vaccinations are strongly recommended to help reduce chance of breathing issues:

- **Flu vaccine** - the flu can be serious, leading to complications such as pneumonia, myocarditis (inflammation of heart), neurologic conditions and other bacterial infections – all high risk to airway stenosis patients.
- **Pneumococcal vaccine** - helps protect you against pneumococcal disease. Pneumococcal can cause a fever, pneumonia, meningitis, blood poisoning (septicaemia), ear infections and other airway infections.
- **Covid-19** – complications from Covid-19 are increased if your airway is narrow, or you have another illness such as diabetes, heart disease or are obese. It is recommended you have this vaccine to minimise the chance of developing severe complications.

Prescription medicine

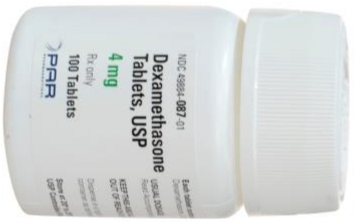
Ideally, your kit at home should include the following:

Antibiotics: You can keep these in the fridge and have them quickly on hand to treat any airway infections. It is important to nip any potential mucus plugs in the bud and having these immediately accessible will help you do that. At the very least have a prescription from your doctor that you can quickly fill.

Prednisone: Inflammation is common with this condition and can cause breathing issues at short notice. Having steroids on hand can help address this quickly.

Make sure the dose is small enough to enable you to taper over a few days. A typical dose would be 20mg for 5 days, followed by 10mg for 5 days, followed by 5mg for 5 days.

We recommend taking additional Calcium and Vitamin D to help support your bone health.



Dexamethasone - corticosteroid medication: An alternative to prednisone steroid tablets, these also help tackle inflammation.

A typical dose would be 4mg three times a day for three to five days.

If available in your country, insist on **enteric coated** steroid tablets. Oral steroids can exacerbate reflux symptoms. Enteric coated tablets prevent absorption in the stomach (which causes or worsens reflux), and instead the tablet travels further and is absorbed in the small intestine.

If they are not available, ensure you take an alginate (seaweed) based anti-reflux medication (such as Gaviscon) to prevent stomach acid reaching your airway and causing more issues.

We recommend taking additional Calcium and Vitamin D to help support your bone health.

Inhaled steroids: A steroid inhaler can quickly deliver steroids right to your stenosis, helping take down localised swelling – a product such as Pulmicort or Flovent will be usually prescribed.

Make sure you gargle and rinse your mouth out with water after each inhale (usually one or two per day) to prevent oral thrush.

Use this when you feel you are narrowing – it can be used in conjunction with a short dose of steroids.

Non-drowsy antihistamine tablets: these are great to have on hand to stop minor allergic reactions – such as sneezing and coughing – in response to an allergen. Anything that helps stop more mucus production and irritation to your airway is a good thing.

Letter from your surgeon for emergencies

If you are travelling and/or do not live near to your treating doctor, it is recommended you ask your surgeon for a letter to have on hand, should you ever need to present to hospital for breathing issues. It should ideally be on your doctor's letterhead with the following message (or similar in their own words).

If you are unable to get this from your doctor, just carrying a printed version of this letter will help in emergency situations.

> Dear Doctor
>
> This patient suffers with a recurrent stenosis of her airway which may lead to shortness of breath or stridor.
>
> She is more likely to have mucous plugging especially in the presence of a respiratory infection. Please avoid a tracheostomy unless lifesaving and do consult with her airway specialist detailed below:
>
> (Doctor's details including emergency contact number)
>
> Given the small airway diameter, intubating the patient may be very difficult, if not impossible. Attempts at intubating the patient without directly visualizing the subglottis can cause further traumatic swelling and worsen the situation.
>
> Consider preventative management until the subglottis can be visualized or the patient can be taken to the operating room to improve the airway.
>
> - **Steroid injection** either IV or IM (Dexamethasone 10 mg IV, Solu-Medrol IV/IM, or equivalent)
> - **Ventilation support** using CPAP, BiPAP, or High Flow O_2 via nasal cannula
> - **Respiratory support** with adjuncts such as Heliox and racemic epinephrine inhalation

Hopefully, you never need to use this, but it is better to have on hand. This letter could save your airway from serious damage.

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 9% normal saline (available from chemists, online or may be provided by your doctor) or cool boiled fresh water (i.e. do not 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 mouthpiece if you do not 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.

What should I use in it?

0.9% ‘Normal’ Saline is recommended for everyday use. This is a mixture of sterile water and salt. The 0.9% means there is 0.9 grams of salt per 100ml of solution, like the amount of salt found in the blood and tears. It may also be known as isotonic saline. It can be purchased, or may be prescribed by your doctor, but it is also very easy to make.

If you have more challenging mucous, you may want to try 3% Hypertonic saline. This is much saltier, and will make you cough harder, but attract more liquid to your stenosis and make the sticky mucous easier to shift. Using the recipe for normal saline, instead of 1 teaspoon of salt, you would make it with 2-3 teaspoons.

Rarely, your doctor may recommend using your nebuliser with a drug called albuterol (also known as salbutamol) in your nebuliser. This is usually used in patients with asthma to help relieve spasms or tightening of the muscles in the lungs and airways. This will need to be on prescription, if used.

Recipe for normal saline

Ingredients:

2 cups of tap water

1 teaspoon of iodine-free salt/fine sea salt/Kosher salt

Method:

1. Bring 2 cups of water to the boil, and boil covered on stove top for 15 minutes (to sterilise the water)
2. Cool to room temperature
3. Mix in salt until dissolved
4. Use as often needed
5. Store in fridge for 24 hours

Why is using a nebuliser useful?

It helps put liquid directly into your trachea, soothing the airway and helping move 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. Nebulising can help you to avoid a life-threatening situation. Patients have died from incurring a mucus plug they were unable to shift.

What types of nebulizer are there?

There are many on the market, but we highly recommend a quiet nebuliser – it means you are more likely to use it, and the two recommended below are also portable and can be used while watching the TV or travelling as a passenger in a car or plane, not disturbing anybody. Two examples used by SGS patients are detailed below:

Omron MicroAir or **Phillips Innospire –**

- 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.
- Run on batteries (last around 4 hours)

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 used them to help young children with croup.

3. Medic alert bracelet

First responders and medical personnel are **trained to first look for medical identification jewellery**. Medical IDs will alert emergency medical professionals to your critical health and personal information and will speak for you in the event you become unresponsive.

Some people have suggested having a tattoo or alternative to jewellery – please note this is **not** recommended as this is not commonplace and they are likely to be overlooked in an emergency.

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 several designs.

You should join the registry of difficult airway/intubation patients which links to your membership.

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

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

Wearing a bracelet reduces treatment errors which may result from not having a patient's health record during an emergency or upon hospital admission.

Suggested wording: Subglottic stenosis; very narrow airway, difficult intubation - use paediatric tube.

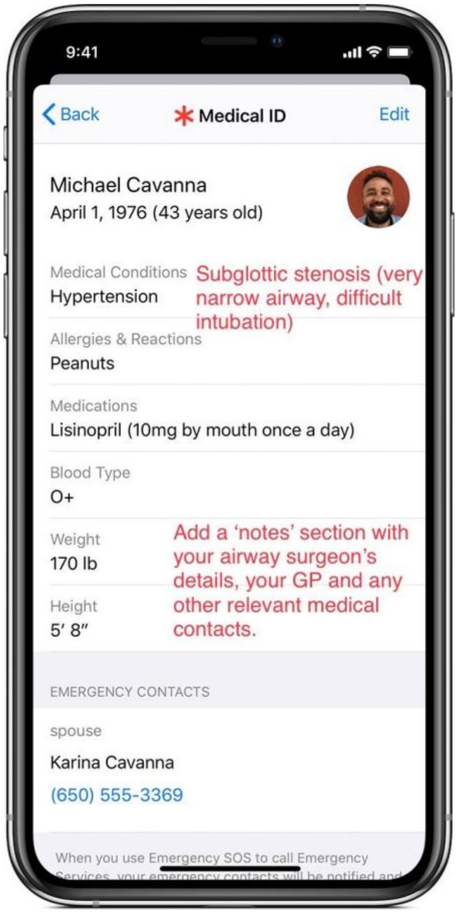
4. iPhone users: *Medical ID

One of the lesser known 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.

How to set up your Medical ID

- Open the Health app ♥ and tap the Summary tab.
- Tap your profile picture in the upper-right corner.
- Under Medical Details, tap Medical ID.
- Tap Edit in the upper-right corner.
- To make your Medical ID available from the Lock screen on your iPhone, turn on Show When Locked. In an emergency, this gives information to people who want to help.
- If you are in the USA, to share your Medical ID with emergency responders, turn on Share During Emergency Call. When you make a call or send a text to emergency services on your iPhone or Apple Watch, your Medical ID will automatically be shared with emergency services
- Enter health information like your date of birth, allergies, and blood type.
- Tap Done.

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



Michael Cavanna
April 1, 1976 (43 years old)

Medical Conditions
Hypertension

Subglottic stenosis (very narrow airway, difficult intubation)

Allergies & Reactions
Peanuts

Medications
Lisinopril (10mg by mouth once a day)

Blood Type
O+

Weight
170 lb

Height
5′ 8″

Add a 'notes' section with your airway surgeon's details, your GP and any other relevant medical contacts.

EMERGENCY CONTACTS

spouse
Karina Cavanna
(650) 555-3369

When you use Emergency SOS to call Emergency Services, your emergency contacts will be notified and

What is a 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 without seeing your doctor for a scope.

It measures your ability to push air out of your lungs in one fast blast by blowing hard through a mouthpiece (i.e., 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 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 to organise injections or other intervention to stem the decline.

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 an app or spreadsheet.

Record the highest of three readings; do not reset the indicator between readings.

How often should I use my meter?

Ideally once a day but at least weekly if you find it hard to remember each day.

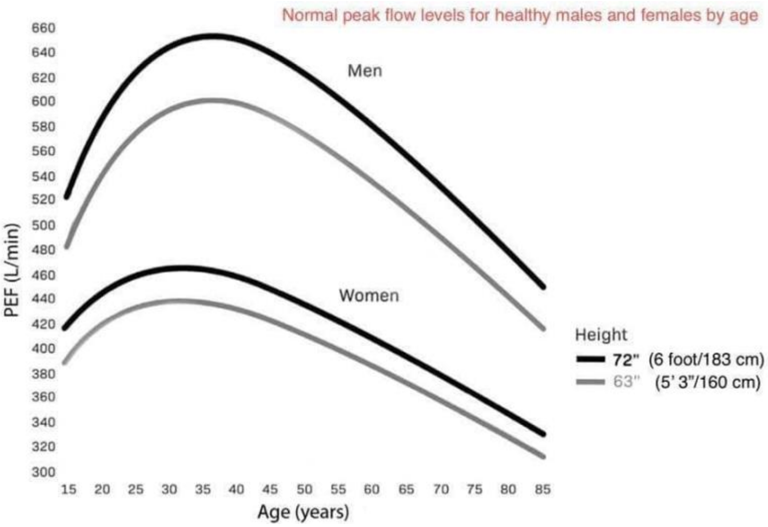
Does it matter if it is not the same time each day?

Try for the same time of day, e.g. when you wake up in the morning – when you clean your teeth. Integrating using your peak flow meter with an existing habit means you are more likely to continue doing it.

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

The data you are recording is individual to you. Everyone has a slightly different expectation depending on their age, height, other health conditions and where their stenosis is located. Men, regardless of their height or age, will always achieve a higher Peak Flow reading than women. It is not useful to compare your result with your brother, boyfriend or husband!

This chart gives you an indication of average normal peak flows for 'healthy' people by age, height, and gender. As you can see, the older you get, the lower your expected PEF is.



Of course, there are other things to consider including other health conditions that may also impact breathing (such as cardiac health, lung conditions), overall fitness. For example, people who maintain a good level of fitness and live at high altitudes can achieve much higher peak flows than expected, whereas someone who has additional health conditions may be expected to achieve a lower level. If you have issues with inflammation and/or mucous, your peak flow is likely to be lower.

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, or you have a mucus plug/inflammation.

Where should I record my values?

You can record PEF results using specifically designed apps – such as the one provided by AsthmaMD (www.asthmamd.org/)

The AsthmaMD app provides green, amber and red zones set by your age and height to help provide a guideline for your airway health. Once you reach close to 'red' it is very likely you need a dilation surgery.

You should contact your surgeon prior to reaching this level to schedule your next procedure.

iPhone users will also find a tracking feature under the Health app which comes with the phone, or you can track it manually on a paper chart, or spreadsheet.

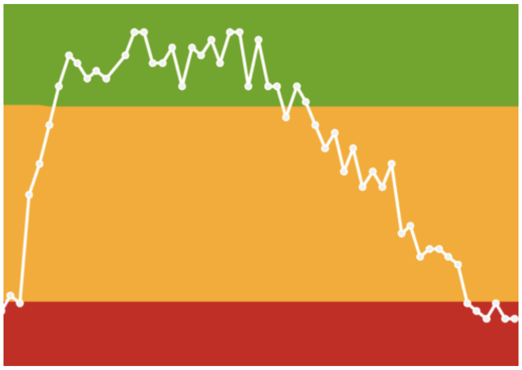


Chart produced by AsthmaMD app

This chart shows a steady decline following a quick increase (post dilation surgery) for example.

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

Resume use of your peak flow meter when it feels comfortable to do so. It is important to understand how good you are post a dilation or steroid injections (when you are breathing well) though, as this will help you learn more about your airway health.

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 breathing well versus less well – you can also monitor how quickly you decline after a dilation or improve after injections.

You can be proactive in making appointments for procedures (e.g a check-up, injections or a dilation surgery) by providing evidence of a continual decline in your airway health.

All evidence is essential to help your specialist offer advice about any potential next steps in your care.

Doctors are very happy when their patients can show evidence of their airway health via a peak flow chart, and there have been many published medical papers in recent years supporting this.

Summary: what interventions are available?

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

Surgical procedures

ENDOSCOPIC (Using a flexible tube, inserted via the nose or mouth)

- Endoscopic tracheal dilation/dilatation (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, anti-reflux)
- Endoscopic tracheal reconstruction (Maddern technique)

OPEN ANTERIOR NECK SURGERY

- Cricotracheal resection
- Laryngotracheoplasty:
  - Cartilage tracheoplasty
  - Slide tracheoplasty
- Tracheostomy (rare)

IMMUNOTHERPY to inhibit immune system response

Your doctor may suggest immune suppressant medication if you have inflammation in your airway, and/or a quickly returning stenosis (6mthly or more often) even if you haven’t tested positive for vasculitis (e.g. GPA). This may include one or more of the following drugs:

- Rituxan/Rituximab
- Mycophenolate mofetil (MMF)/ Cellcept
- Methotrexate (MTX)
- Azathioprine/Imuran
- Cyclophosphamide/Cytoxan

Medical therapies

ANTIBIOTICS to tackle bacteria which may be causing inflammation

- Bactrim, a sulpha drug
- Azithromycin
- Erythromycin, if allergic to sulpha drugs

CORTICOSTEROIDS to inhibit fibroblast formation and reduce inflammation

- Oral tablet (e.g. prednisone)
- Oral inhalation (e.g.nebulized albuterol, budesonide, eg., pulmicort)
- Injected (e.g., triamcinolone, i.e. Kenalog, between dilations in-office or "awake")

PROTON-PUMP INHIBITORS (PPIs) to reduce gastric acid reflux (GERD) if this is an issue for you’.

It is not recommended PPIs are taken for longer than 6 months.

Please talk to your doctor if you have been taking these for longer – research has shown prolonged use leads to decreased intestinal absorption of calcium resulting in negative calcium balance, increased osteoporosis, development of secondary hyperparathyroidism, increased bone loss and increased fractures.

Sodium alginate liquid

Note:
This information has been compiled from published journal articles, physicians’ recommendations, and personal communications. It presents SGS testing, therapy or monitoring options that patients may be following. It does not intend to endorse or repudiate any SGS testing, therapy or monitoring options.

Complementary therapies

HYDRATION to keep mucus thin and slippery (not thick and sticky) and to keep vocal folds moist

- Internal hydration:
  - Drink plenty of water
  - Reduce caffeine and alcohol
- External hydration:
  - Steam inhalation
  - Nebulisation with sodium chloride solution
  - Warm air room humidification
  - Nasal rinse
  - Nasal spray

MUCOLYTIC THERAPY to help expel mucus

Mucolytic medication:

- OTC expectorant cough syrup (e.g., guaifenesin)
- Throat lozenge with glycerine, pectin, or slippery elm (not menthol or eucalyptus)
- Amino acid (N-Acetyl Cysteine, NAC):
  - OTC oral capsule
  - Oral inhalation (nebulized NAC, i.e., Mucomyst)

Airway Clearance Technique or Device:

- ‘Huff cough or ‘’huffing exercise
- Flutter valve, e.g., Acapella

LIFESTYLE CHANGES

- Dietary change, anti-inflammatory diet, anti-reflux diet
- Weight loss
- Vitamins, herbs
- Probiotics
- Enzymes (e.g. bromelain)

What is a dilation surgery?

Once you have been diagnosed with SGS your surgeon will most likely book you in for a dilation operation (a dilation and a dilatation refer to the same thing – an operation aimed at opening your narrowing). 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 different types of dilation. What is the difference between these?

Technique	***Pros***	***Cons***
Laser dilation (or laser and balloon or laser and manual manipulation of scar)	A precise manner of dilation, particularly in experienced hands allowing the doctor to carefully open the scarring without damage to the rest of the airway	In less experienced hands there is some risk of the laser causing damage to healthy tissue therefore making the scar tissue spread.
Balloon dilation (sometimes with a small blade to do some cuts first)	Not likely to cause problems in other areas of the airway/cause the tissue to grow.	Slightly less precise, using the balloon to 'tear' open the scar tissue. If cuts are done first, then the balloon can open the scar tissue in a more regulated way.
Coblation dilation (Radio-frequency cold coblation) - uses controlled energy delivery to operate at the relatively low temperature of 60-65°C.	This aims to reduce surrounding tissue damage and postoperative pain and to avoid seeding of viral particles during the process.	Even in experienced hands it can be a challenge to control the exact area being treated. Anecdotal findings from doctors suggest there is a risk of this method causing the scar to travel up towards the vocal cords meaning it is not recommended.
Cryotherapy (could be cryotherapy with balloon). This uses a technique whereby the scar tissue is frozen and defrosted before being dilated.	There is limited research suggesting this can be successful among some patients	Not used in the major centres. Key concerns are the inability to direct the cryotherapy only to the scar tissue, opening the healthy airway tissue surrounding it to potentially becoming damaged and scarring. This is thought to cause the scar to travel up the airway closer to the vocal cords (thereby causing permanent loss of voice).

Technique	***Pros***	***Cons***
Rigid bronch (use of a set of steel pipes, small to larger, in succession, to stretch the trachea) without balloon.	Doctors with a lot of experience with this technique prefer it to laser or balloon and believe it delivers better results.	The surgical view is quite poor with this technique resulting in a less precise procedure. High risk of permanent damage to vocal cords and therefore voice.
CO_2 laser wedge excision (sometimes referred to as wedge resection)	Using a laser, the doctor will cut wedges out of the scar tissue. The patient will take some medication post-surgery to maximise success rate.	This technique (and associated medication protocol) has high success rates in keeping the airway open while minimising risk to vocal cords.
Argon Plasma Coagulation (APC) - an electrosurgery using flexible endoscopy to treat trachea/bronchial stenosis. APC uses gas discharges to induce thermal therapy with small electrical bolts on soft tissue with no contact, meaning no cutting. This allows the surgeon to carefully destroy the scar tissue.	Minimum bleeding from stenosis removal. Less invasive option for treating stenosis. May be beneficial in removing more difficult scar tissue.	Some surgeons like to perform a series of this procedure (3-4 times every few weeks) to arrive at the best outcome. Whether this is more effective than a single procedure, is uncertain.

Use this information to **ask questions before you have your surgery**. Make sure you know what operation you are having in advance. If you have had one of the 'higher risk' dilations in the past, do not panic - it does not mean your scar tissue is going to travel up towards your vocal cords, but you may want to ensure you have a less risky surgery in the future.

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?

'Jet ventilation' is the most used technique. This allows continuous oxygenation during the dilatation procedure without intubation.

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 sometimes 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 little 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). Some patients experience a longer time between dilations when this is applied. Use of mitomycin C seems to be declining with most experienced surgeons.

My Doctor mentioned steroid injections with my dilation – what does this mean?

It is becoming increasingly best-practice for steroids to be injected at the site of your scarring once the dilation has been completed. This will be in similar quantities to those injected in office (see section on in-office/awake steroid injections) but administered while you are under a general anaesthetic. It is possible you may experience minor side effects (such as flushing and/or insomnia) from these for the 24 hours following your surgery.

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 two or three days you should feel the benefit breathing wise.

Some people may have a husky voice for a few days post operation but this will go as any bruising and inflammation subsides.

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 be sore. You may also feel aching in your ribs from the air flow in your lungs. All this dies down within a couple of days.

There is a nerve in the airway which links directly to your ear. As this may be irritated during the surgery, there is a chance you will have pain in one or both ears for a short time (up to a week).

You may have a cough for a week or so in response to the healing. This is normal.

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

What medication am I likely to be on afterwards?

Some doctors prescribe Tylenol 2 with Codeine (USA) or Panadeine 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.

If you have the endoscopic laser wedge excision/resection you are likely to be asked to use inhaled corticosteroid and trimethoprim/sulfamethoxazole (an antibiotic such as Bactrim, Cotrim or Septra) post the procedure. Some patients are also prescribed antibiotics for 5 days post-surgery.

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. Do not whisper as this strains the vocal cords.

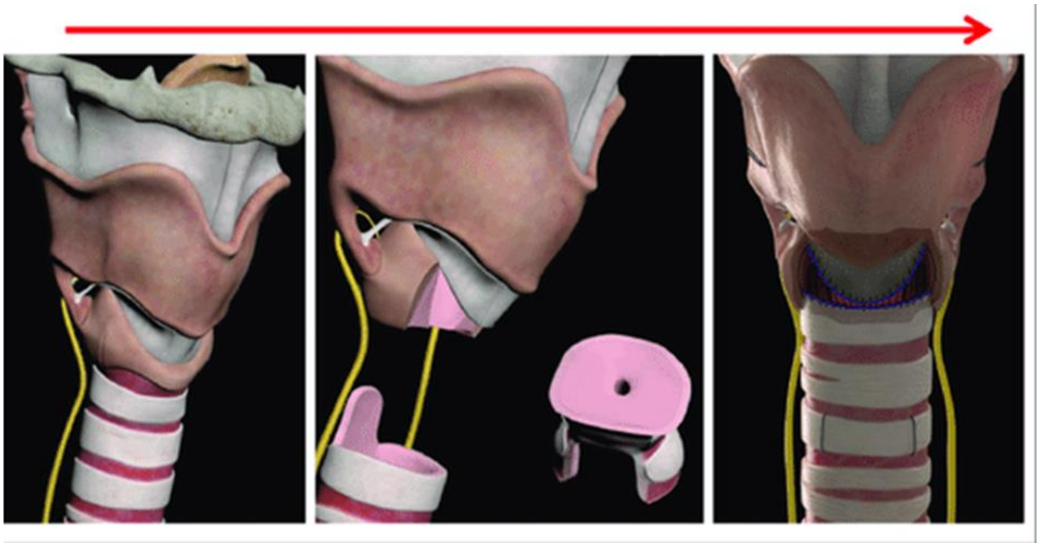
How long until I can resume normal activity or exercise?

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 would not 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 do not feel like it, then don't do it! Do not 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

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. You may hear it also called Cricotracheal resection (CTR), Laryngeal Tracheal Resection (LTR) or Tracheal resection and reanastamosis. All are slightly different but with similarities. For example, when the scarred portion is particularly long, 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. On average, a resection will last around 10 years. As with all averages, some last less time, others longer.

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

Most 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?

The main risk is restenosis. Next is damage to the nerves of the vocal cords. There is about 1% chance of bleeding or infection. Around 5% of patients with a very experienced surgeon will go on to re-stenose within the immediate 3 years.

Most patients will have some permanent change in voice (often deeper) due to changes in laryngeal elevation (needed for a good vocal range, especially in singers). Some may not be able to project their voice or speak loudly.

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 (if used) 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 can be used to help keep the head in position in the case of long segment stenosis removal, but the trend has generally to be to move away from this technique. If your surgeon plans to use a chin stitch (sometimes called a 'Grillo stitch' after the first doctor who used this technique) **please ensure you are comfortable with their expertise and their reasons for using it.**

Doctors who have been performing many resections with the chin stitch for many years are confident in the safety of this technique. If you are certain with the level of experience of your surgeon, then you should be safe with this technique.

These days, it is more common to have a c-collar for a few weeks and keep the neck flexed as much as possible.

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

Most patients 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 feeding tube and catheter? If so, for how long?

You are likely to be given catheter. If 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.

Are you required to be on voice rest 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 does it take for your voice to get as good as it will get?

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

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

Generally a minimum of 6 weeks off work, and return to work depends on each individual, as everyone feels and recuperates differently. Many patients are told they cannot drive for 8 weeks

How bad is the scar?

The scar is about 7.5cm (3 inches) in length.
Can also have a small 1 cm (half an inch) scar where the drain is located

What is The Maddern Procedure (Endoscopic Resection and Mucosal Reconstitution with Epidermal Grafting)?

This surgery is suitable for patients with an isolated subglottic stenosis in a mature, non-inflammatory phase. Through an endoscopic approach (transoral – i.e., via the mouth), the stenosis is completely removed with the assistance of a soft-tissue shaver. The cricoid is then relined with either a buccal or skin graft, held in place for 2 weeks with a temporary silicone stent. This technique is minimally invasive, has the potential to preserve voice function completely, and may have a curative outcome.

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 cords 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, nose 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 may be 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 do not 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 (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.

When can I eat and talk normally?

Once a speech and language 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 your airway is smoothed.

The graft site on your leg, nose or inner cheek will be the sorest point, so keep this covered and protected. It is 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.

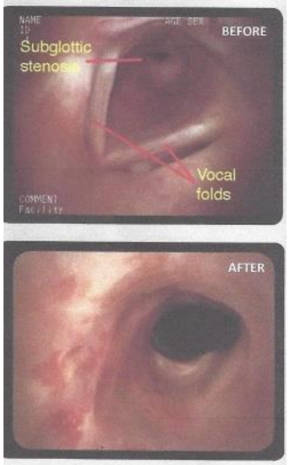
How long does it last?

The first patient to have this operation in 2012 is still breathing well with no restenosis.

Latest research suggests 80% of patients continue to breathe well with no restenosis for at least 5 years.

What are in office/awake steroid injections

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



he above photographs show a patient's subglottic region efore and after treatment with steroid injection.

*Image source – Harvard Otolaryngology 10/2014

Why would I choose to have a steroid injection?

Inflammation is a key part of SGS 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 set of injections 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 four to six weeks for a course of 4-6 injections.

How long does the procedure take?

Including the local anaesthetic applied to your vocal cords and trachea, and the procedure itself, you should expect to be in the office for between 30 and 60 minutes.

Am I sedated or put under a general anaesthetic?

There is no general anaesthetic involved nor sedation (unless you specifically request it). 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 bronchoscope 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, also the thickness of your scar and the ease of penetrating it with the needle.

Will it affect my voice?

Some have a short-term huskiness while the vocal cords 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, often with higher peak flow numbers than they get with a dilation.

What are the risks of steroid injections?

Recent research (2022) finds 65% of patients receiving injections encounter temporary side effects with 23% experiencing 3 or more. These included flushing (38%), insomnia (23%), injection site pain (22%), temporarily increased stridor (20%), coughing up small amounts of blood (16%), heart palpitations (15%), fatigue (10%) and blurry vision (1.9%). Side effects requiring additional medical care and/or treatment are rare.

What is the Mayo Protocol?

Several patients are following this regimen, often with good results. You might like to ask your doctor about trying this, or perhaps they will suggest it to you.

The protocol involves taking three drugs:

- Trimethoprim/Sulfamethoxazole (Bactrim/Septra/Co-Trimoxazole) – 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 cords (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. Recent research suggests PPIs do not have an impact on patients' airways where there is no evidence of GERD. A gentler medication such as Gaviscon may be better in this situation.

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.

How can immunotherapy help my subglottic stenosis?

Immunotherapy is becoming a more frequently used approach for patients who are having frequent dilations – by frequent we mean every six months or more often. There are a number of options available, and commonly used for other health conditions, so the side effects are well known and they are not considered experimental.

Rituximab is the most frequently used (also known by its brand names of MabThera, Rixathon and Truxima), but if this does not work successfully for you, there are other options available such as CellCept (mycophenolate mofetil) which also works to help your body stop attacking what it perceives to be a foreign body (such as your airway) by inhibiting B-cell and T-cell function. This chapter concentrates on Rituximab as the most used immunotherapy drug for airway stenosis.

What is Rituximab?

Rituximab is a drug most used to treat severe rheumatoid arthritis, as well as diagnosed autoimmune diseases such as granulomatosis with polyangiitis (Wegener's), also known as GPA.

But I haven’t been diagnosed with GPA, rheumatoid arthritis or a known autoimmune disease – why would I have this treatment?

There is a well-documented train of thought that suggests ‘idiopathic’ subglottic stenosis is a localised form of GPA and therefore treating it with the same medication will have good results despite biopsies and blood tests not proving GPA is present (you might hear you are ANCA negative). Even if you are ANCA positive, biopsies may not detect any signs of GPA.

What does Rituximab do?

Rituximab targets white blood cells in your body called B-cells (B-lymphocytes). These cells are part of the body’s immune system, responsible for attacking what they perceive as foreign bodies, and for creating an inflammatory response. Inflammation is frequently noted by doctors treating subglottic stenosis.

Is this a cure for subglottic stenosis?

For many patients, infusions of Rituximab put subglottic stenosis into remission, meaning it stops the airway closing up. Most patients get a longer period surgery-free, with studies frequently stating an average of 31-32 months (more than 2.5 years) between dilations. Your doctor will monitor your reaction closely and advise if another course of Rituximab is recommended.

How do patients receive this medication?

It is administered via an intravenous infusion – straight into a vein in the back of your hand or in the crook of your arm.

The infusion takes around 6-8 hours as it is given to you slowly, allowing medical professionals to monitor your reaction to the drug and act quickly to slow or stop administration if you have any adverse reactions.

Before the Rituximab is administered, antihistamine and steroids will be infused. This is to help minimise any adverse reactions to the drug.

So, is Rituximab chemotherapy?

No, it is not chemotherapy. It is a targeted antibody therapy – immunotherapy. It can be used alongside chemotherapy with cancer patients, but is not chemotherapy itself.

How often do you have to do it?

The initial treatment usually consists of two to three sessions (about 6 to 8 hours each) two weeks apart. This will then subdue your B-cells for approximately six months. Depending on how well your stenosis responds (i.e. whether you need another dilation) you may need another round of two sessions 6-10 months after your first. Your blood will be monitored regularly. Patients who are positive for GPA (with symptoms impacting other areas of their body in addition to their airway) may need four sessions.

How long does it take to make a difference?

It takes about six weeks to take its full effect.

Do you need to take any other medication?

Some doctors prescribe prednisone to be taken orally for several months, while others may prescribe a low dose antibiotic to help prevent any bacterial infections, taken long term.

If I have a limited immune system, am I at risk from other illnesses?

If you catch a virus, such as Covid-19, a cold or flu for example, you are likely to get sicker for longer, and more severely than you would if you did not have this treatment. It is advisable you are vaccinated against Covid-19 (and potentially pneumonia, shingles and flu) before starting infusions – talk to your specialist about their recommendations. Blood tests will check you have not had hepatitis-B and assess the current health of your blood, lungs and kidneys. People with existing serious infections should not receive Rituximab.

How should I prepare for an infusion?

- **Remain hydrated** - particularly the day and evening before your infusion, which will help in finding a vein on the day.
- **Avoid caffeine on the morning of your infusion** - it can raise your blood pressure and constrict your veins
- **Wear comfortable clothes and shoes** – you will be attached to an IV pole so it will be challenging to remove clothing if you are too hot, and using the bathroom will require the use

of only one hand. No complicated trousers or underwear! There are usually blankets if you get cold, but if you have long sleeves you won't be able to remove a top to cool down. Ideally wear or bring footwear that is easy to slip on and off.

- **Move every hour** – you are sitting for a long while. If you are keeping hydrated, you should be needing those bathroom breaks, and take advantage of it to avoid pressure sores and risk of clots.
- **Bring easy to eat snacks** – again, you are one handed, so any food or drink you bring with you should be accessible
- **Bring phone with music and headphones** – the drugs (particularly the antihistamine) may make you feel drowsy, so you are unlikely to want to read. Napping and listening to music is good.
- **Don't forget your charger** – you're there for a few hours, so make sure your devices stay working

How will I feel during the infusion?

You are likely to feel drowsy. You will be sitting in a chair with your blood pressure being regularly monitored on one arm, with the infusion entering a vein on the other. If you have any reaction at all, no matter how minor you think it is, let the attending nurse know. This may include feeling itchy, numb lips, tingling tongue.

There will be a lot of beeping throughout the infusion. Normally this is to remind the nurse to come and check your blood pressure, and speed up (or slow down) the infusion. It will also beep if the line is bent or obstructed.

How will I feel afterwards?

Prevention is better than cure, and ensuring you are well hydrated should help you avoid the headache which plagues some people. Fatigue is the most common side effect, with some people feeling as though they have flu-like symptoms for up to four days.

You are given steroids intravenously which can cause insomnia for a night or two, so despite feeling tired, you may not be able to sleep. You may feel a temporary hunger (from the steroids), and other short term side effects are a burning sensation to face and skin and a dry, scratchy throat.

Can I still have other treatment on my stenosis as well as this?

Yes, all usual options (eg steroid injections/dilation) remain open to you while on this treatment plan.

What questions should I ask my surgeon?

At your **very first appointment** with your doctor these questions will help you determine whether this is the right doctor to be treating you. You only get one airway and you want it handled correctly from the start:

How much experience do you have with airway stenosis?	*Establish how confident the doctor is in this field, given it is a rare area of treatment and many ENTs will have only read about it while studying. The more patients they treat and the longer they have been involved with airway stenosis, the better – you do not want to be their first patient.*
How many airway stenosis patients do you have?	
How long have you been treating patients with airway stenosis?	
Where/with whom did you do your training in this area?	*If they trained at a high-volume centre then it is likely they have a lot more experience and therefore you can feel confident they have learnt from the best.*
What is the average reoccurrence rate of your patients' stenoses? i.e., how frequently do your patients generally have to return for a dilation surgery?	*Alarm bells should ring if their patients are all returning for surgery every six months or more frequently. This should ideally be the exception rather than the rule.*
Has any of your airway stenosis patients ever needed to have a tracheotomy? **Under what circumstances?** **What about other complications?**	*Tracheostomy is a rare (but sometimes necessary) occurrence among airway stenosis patients – important to understand what led to this (or other complications) happening.*
What are your thoughts regarding putting a permanent stent into my airway to keep it open?	*This is a trick question – any doctor with experience in this area would NOT recommend a stent – there are many risks – including complete blockage (requiring a tracheotomy), movement of the stent and creation of more scar tissue, movement and damage to vocal cords. If your doctor entertains this as an option, seek another opinion.*

What treatment options do you offer? e.g. steroid injections/dilation/wedge resection/Maddern/resection **How many dilation surgeries have you carried out in the past three months?**	*The treatments offered (and variety of them) will help you understand their experience. More experienced doctors/centres offer more options (or will be happy to refer you to another surgeon and know who to send you to). The more surgeries they have carried out in the past three months, you can infer their skills will be more up to date.* *If your doctor does one thing well (eg dilations) that is great, as long as they are aware of other treatments and where to send you, should you choose that path.*
Where would you refer more challenging patients to, if anywhere?	
What do you propose to be the plan for my ongoing treatment?	
What do I do if my breathing suddenly deteriorates, and I need to see you urgently?	*As an airway patient, the majority of doctors will put a flag on your record to alert their gatekeeper (admin/appointments/secretary) to prioritise your appointments if you need one urgently. It is worth raising this with your surgeon to make sure it is the case so you don't have issues scheduling an appointment. They may also recommend particular hospitals to attend/avoid if you need to present at emergency.*
Please can I have a copy of the image of my stenosis?	*The doctor should look at your airway using a laryngoscope/bronchoscope which involves a camera going up your nose and down your throat. Usually the doctor is able to sight your stenosis through your vocal cords, and should be able to save a photograph of your stenosis.* *If you are able to take a printed copy of this image it gives you something to take to other doctors, should you choose to go elsewhere.*

When having **dilation surgery:**

How long should I expect to be in hospital?	*Most dilation surgeries are done as day surgery, but if you are susceptible to swelling, have other potential health issues or have a long way to travel then you may need to stay overnight – either in hospital or within easy reach of the hospital.*
What can I do to protect my teeth from damage?	*Many hospitals will insert a mouthguard to protect your teeth from the surgical instruments, but not all. It may be worth getting something specific to fit your teeth, if your surgeon is agreeable to this.*
What surgery are you planning to perform? A laser dilation? Balloon? Coblation?	*Understanding what type of dilation your surgeon plans to proceed with is important. For example, coblation is a dilation technique performed at several centres, but anecdotally is not recommended as it increases the likelihood of the scar spreading up your trachea. If your surgeon suggests this technique, then request an alternative or seek a second opinion with a surgeon that does not do this operation.*
Will you be doing any steroid injections? Applying Mitomycin-C?	*Being aware of what treatment you are having is important in assessing success rates, side effects and so on, especially if you may potentially see another surgeon at some point.*
Are you planning to do a biopsy of the scar to test for ANCA? Or take bloods to test for this?	*A test for ANCA (autoimmune disease) is recommended each time you have a dilation. A negative result in the past does not mean there might not be a positive result in the future.*

At **every appointment** with your specialist:

What is the current diameter of my stenosis in millimetres?	*Your doctor can best answer this when they have seen your airway under a general anaesthetic, and they can measure it accurately. This will be in millimetres (mm).*

At which percentage is my airway closed?	*This is going to be a best guess by your doctor when you are seen with a scope. If you know the diameter of your airway when it is at its most open, you can calculate the approximate diameter in millimetres (mm).*
What is the current distance of the top of my stenosis from my vocal cords?	*This will help you to understand any risk to your voice quality from the movement of the scar and may help you make decisions about a change to the type of treatment or surgery you receive.*
How has my stenosis changed since my last appointment?	*You are aiming to understand what is happening in your airway – whether the scar is changing – is it moving towards your vocal cords? Is it moving further down your trachea? Is the scar getting thicker or thinner? Is it corkscrewing or remaining the same? Are there any new areas of scarring?* *Your surgeon should take regular photos and /or have a detailed description of the stenosis each time to refer to and make comparisons with previous exams.*
What are my ANCA test results? And when were they last tested?	*If you have been diagnosed idiopathic (not if you have been diagnosed with GPA or other autoimmune (AI) disease) then it is recommended this is tested annually along with a basic-metabolic profile – your GP/primary care doctor can usually organise these. Even if you have tested negative in the past, the result can still change, and the treatment will be slightly different if you are shown to have an AI disease.*
Do both my vocal cords work as they should?	*If there are any issues with your voice it is worth understanding whether it is related to your scar, to mucus or whether the vocal cords are not working properly. Knowing this will help you to get a referral to a specialist to help with voice such as a speech and language therapist or otolaryngologist which specialises in voice.*
Are you aware of any clinical trials or studies I could be a part of?	

When considering **major surgery:**

How many resections/reconstructions have you done? How many on idiopathic patients? How many on non-idiopathic?	*Help understand your surgeon's experience in this difficult and delicate surgery. You want a doctor who has done many successful operations, specifically with patients like you. You also want one with an excellent team around them and a recovery department at the hospital which is used to dealing with this type of surgery.*
What is your success rate? How do you define success?	*For example, a resection will last, on average, ten years. Does your surgeon consider this a success? What percentage of their patients return for more surgery within a decade?*
What do you see as the general risks of this surgery?	*For example Restenosis? Nerve damage? Infection? Trach? Weak voice? Unable to sing?*
What do you see as my personal risks?	*Help you understand any potential issues specific to your situation and health – also help you feel comfortable the surgeon has considered all options in recommending this operation for you.*
How can I personally reduce my risks of complications?	*For example this may mean gaining or losing weight, cutting out bad habits, doing pre-operative exercises or stretches.*
How long will I stay in the hospital?	*You may have work and/or family issues to consider when booking this surgery, so timing can be a consideration in deciding whether to or when to proceed.*
Can I talk to any of your former patients?	*Most experienced surgeons will have patients who are willing to share their surgery story and help you decide whether you are comfortable proceeding with the surgeon and procedure.*

What is your major complication rate? What major complications have you experienced?	*This will both help you prepare for the worst-case scenario, and help you build confidence in your surgeon's ability to cope with unexpected issues during a major surgery. If you feel uncomfortable with their answer, then seek a second opinion.*
What happens during the operation? How long does it last?	*The more you understand in advance, the more comfortable you will feel about the surgery.*
What can I expect when I wake up? (e.g. lines, drains, masks, catheter, feeding tube)	*If you know what to expect when you wake, then you will be mentally prepared to accept your situation and move forwards with healing. You can warn friends and family in advance what to expect if they visit you post-surgery.*
How will I feel when I wake up? (Pain? If so where)	*Understand more about managing expectations and how the recovery team will be managing this – through pain killers or muscle relaxers, steroids and so on*
When will I be able to eat and drink normally?	*For example moving on from ice chips to solid food or having a feeding tube removed.*
How long before I can go home?	*Understand how long you are likely to be in hospital before discharge. If you live a long way away from the doctor and need follow up appointments, it may mean staying somewhere nearby to attend those.*
What sorts of complications might I encounter after leaving the hospital and how should I deal with those if they occur?	*It is worth having a plan in place for any difficulties such as challenges breathing, swallowing, or talking, pain in your neck or elsewhere, and how to manage those. Will your doctor share a direct email or phone number for you to contact? Or is there a nurse or registrar you can call if you are concerned?*
Will I have any restrictions once I go home?	*For example there may be restrictions on voice (voice rest), driving or exercise. You may need to eat soft food for a while or avoid certain foods or drinks.*

Am I likely to need to take medication once I am discharged? For how long?	*These could include steroids, anti-reflux medication or antibiotics for example.*
Is there any equipment I would need to use at home?	*For example after surgery you may be told to nebulise three times a day to help keep your airway moist and aid the healing. Others may recommend a humidifier to make dry air (e.g. in heating or air conditioning) easier to breathe.*

Above all, make sure you never feel bullied into having surgery with a doctor if you do not feel 100% comfortable. It is your right to seek a second opinion, it is your body, you can always say no.

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!).

Don't forget to pack your patience ...your surgery may have to be rescheduled due to unforeseen circumstances (such as someone who is a higher emergency than you or complications in someone else's operation).

Going in for a day or one night

If you are just planning to go for one day, we would recommend being prepared to spend the night, in case there is some unexpected inflammation or your surgery is late in the day and they are not comfortable with you leaving.

Information

- A copy of your latest peak flow chart (or show it via your app) to show the doctor and anaesthetist when they visit you pre-op – help them be prepared for how your airway might look once you are under an anaesthetic
- Pen and paper or whiteboard in case you need to communicate without using your voice
- Notebook to jot down any info shared with you post op (or be prepared to use the notes in your phone)

Medical essentials

- Reading glasses and case
- A list of all your medications (this can be provided by your pharmacy or Doctor surgery)
    - Bring in 3 days' worth (better to be safe than sorry)
- A list of any herbal remedies and vitamins you are taking regularly
- A note with details of any other medical conditions including details of the medical professional(s) treating you
- Details of any allergies
- Your medical healthcare number and/or health insurance details
- You may also want to bring lozenges for the inevitable sore throat

Clothing

- Wear big knickers, as these will be your only clothes ;) (Apart from those you arrive and leave in of course!)

  - If you are afraid of leakages from coughing fits, then pads
- Flip flops or slippers for walking around on cold tiled floors
- Night dress or pyjamas (short sleeves in case you have a drip/cannula)
- A dressing gown or cardigan you can wear over your gown to keep you warm

Toiletries

- Lip balm as the air conditioning is quite dry (make sure it is clear as a tinted one will end up smeared all over your face - not a good look!)
- Hand cream
- Wash (toiletries) bag – you will want to clean your teeth, possibly your own soap
- Tissues
- Night-time routine things - face creams etc
- Eye mask and ear plugs – Bluetooth sleep headphones with inbuilt eye mask have been recommended
- Hair tie and brush/comb
- Rechargeable fan or hot water bottle – depending on the weather!

Entertainment and communications

- Book or tablet for killing time while you wait – games, music etc
- Phone with internet connection so you can keep your 'Living with idiopathic subglottic stenosis' group friends updated and they can keep you company ;)
- Laptop/tablet (especially if the hospital has free Wi-Fi or you can use your phone data!)
- Don't forget your charger with an extra-long lead – the wall plug is usually a long way from the bed and/or a spare battery pack
- Headphones
- Mini speaker if you have a private room

Food and drink

- Any favourite snack that you can cope with eating rather than the hospital food once you wake up
- Your preferred coffee/tea bags

Going in longer term (e.g. 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
- Load your tablet or laptop with movies and music - bring your headphones
- 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 and neck
- Sore muscle rub/balm. e.g. Icy/Hot.
- Extension lead with multiple power points for plugging in your laptop/tablet/phone/etc
- Microfibre hair turban and travel hair dryer – for when you have the luxury of showering and washing your hair

Tips to deal with major airway surgery in hospital

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

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 a form of fasting diet (e.g. 5:2 or 16:8 – there is a lot of information on the internet about these ways of eating) for some time 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.

Be prepared – ask your doctor about the procedure, talk to other patients who have gone through the same thing, read blogs and books by patients (for airway reconstruction, a good one is: https://dl.bookfunnel.com/kw5xarp4oc)

Try to recognize progress every day, however small it is. It will motivate and encourage you - e.g. - 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 can 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.

Establish your own routines - This is about gaining a bit of your own power rather than being solely at the mercy of the hospital system e.g. 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.

Communicate - not talking is quite disturbing especially if you can't. People treat you differently and try to assume what you are thinking or trying to communicate. Work out the best way to communicate. E.g. use a Bluetooth keyboard with a tablet - have the tablet on view to your guest/nurse and type away all the things you are feeling and thinking or use a white board and marker.

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 are also woken for pain meds and blood pressure and so on.

Having some noise cancelling earphones is essential – e.g. 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. 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. You may decide your continuous sleep more than your 1am pain meds – for example organise that you press your button the moment you awake with pain. That can bring you nearly six hours of continuous sleep - absolute luxury!

Take control of your treatment. Understand what you are being given and why. Again this is about empowerment and 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 am having non-airway surgery – what should I tell my surgeon?

If you must 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 anaesthesiologist 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.

Here is an example email you may want to use to contact your anaesthetist prior to your surgery. Copy your airway surgeon and your non-airway surgeon to the message:

Dear Dr *[insert name]*

I am having an operation with Dr *[name]* on *[date]* and I understand you are to be my anaesthetist.

I am writing to make you aware that I have Idiopathic Subglottic Stenosis. I have attached a document which explains more about the disease and how it impacts patients' airways. *[You have permission to share this Rough Guide]*

Ideally, I will not be intubated for this surgery - if possible, I would prefer a Laryngeal Mask Airway. If I do need to be intubated, I respectfully request:

- Intubation with a small tube - preferably no larger than 6.0mm *[smaller if your airway doctor recommends]*
- ETT cuff pressure measurements (suggested 20-30 cm H_2O pressure?)
- Glide-scope / fibre-optic intubation, if available.

My breathing is good currently with a peak flow of *[insert latest PEF number].*

Of course, I defer to your professional expertise in providing me with the safest care possible.

Many thanks, and I look forward to meeting you. If you would like to talk to me prior to *[surgery date]*, my phone number is *[insert number].*

Kind regards…

Pregnancy and stenosis

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

For reasons unknown, pregnancy seems to be a trigger for some patients with SGS. Given that most patients affected by the disease are women, oestrogen is thought to play a role in the development of SGS. Unsurprisingly, oestrogen 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 the person who will be monitoring your pregnancy about your stenosis.

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

Even if you do not 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 necessarily 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 SGS patients can be treated in the office with awake, unsedated 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 (i.e. intravenous or oral) and inhaled steroids (both nasal and pulmonary) have an established track record in pregnancy (particularly is 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 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 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 anaesthetic drugs can theoretically cross the placenta. Yet despite years of animal studies and observational studies in humans, no aesthetic drug has been shown to be clearly dangerous to the human foetus. There is no single optimal anaesthetic 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 necessary?

While this may be necessary in some cases, the majority of with SGS 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 SGS - do I need a C-section, or can I deliver the baby naturally?

Many SGS patients have delivered babies naturally, so stenosis alone should not necessarily be a reason why you cannot 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 SGS 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.

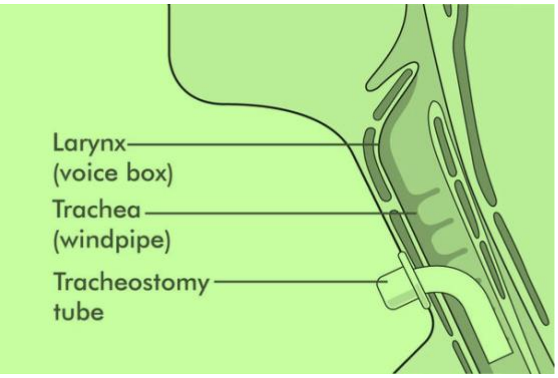
Try to relax! While it can be stressful and scary dealing with this condition during pregnancy - try not to get worried about it.

Tracheostomy – your questions answered

Most patients will not require a tracheotomy and one should not be given to you without prior discussion with your consulting surgeon unless it is deemed potentially lifesaving by the local airway (otolaryngology/anaesthetic/ER consultant) team i.e. an emergency.

However, occasionally patients with airway stenosis need to have a tracheostomy to help them breathe. The thought of this can often feel quite frightening. Hopefully, we can help you feel more at ease.

What is a tracheostomy?



Tracheostomy is a hole that surgeons make through the front of the neck and into the windpipe (trachea). A tracheostomy tube is placed into the hole to keep it open for breathing. The term for the surgical procedure to create this opening is tracheotomy.

Why is a tracheotomy performed?

A tracheotomy is used to help deliver oxygen to the lungs if a person is unable to breathe normally. If you cannot breathe normally it is known as respiratory failure.

A tracheotomy can also be used to bypass an airway that is blocked due to obstruction such as the scar tissue you have as part of your stenosis, swelling, or a mucus plug.

Tracheotomy is most frequently planned and carried out in a hospital environment. But occasionally in the event of an emergency a tracheotomy will be performed outside of a hospital for example the scene of an accident.

Are all airway stenosis patients given a tracheostomy?

No, most patients will never have one. A recent survey amongst airway stenosis patients found that 79% of stenosis patients have never had a tracheostomy, with around one in three of those that have (32%) being temporary (for a month or less), often just as part of a surgery.

How can I avoid needing a tracheostomy?

Sometimes it is unavoidable if part of a major surgery and required as a temporary measure in the event of inflammation. However, on a day-to-day basis there are several things you can do:

- **Monitor your airway** with a peak flow meter so you know when your airway is approaching its next dilation or steroid injection. Do not leave it too long before booking in for your next treatment (operation or injection) – talk to your doctor to ensure you understand the lead time required to book in for a procedure (it may be longer than in the past due to Covid-19 restrictions)
- **Regularly nebulise** and ensure you are hydrated (water, not caffeinated, sugary or alcoholic drinks) to help avoid mucus plugs

- **Have your emergency medical kit** available at home – if you notice you have an infection or are struggling to breathe, be prepared to take antibiotics and/or a short course of steroids to see you through until you can see your doctor in person
- **Take your emergency letter with you (see resources)** - If you must go to hospital in an emergency for your breathing, ensure you take along the letter from your surgeon (template on page 24)

If you are advised that you need to have a tracheostomy - do not panic.

Is a tracheostomy painful?

Any pain should be short term, usually for the first week after its insertion. Any pain should soon settle down and pain relief can be prescribed by the doctors caring for you in hospital.

There can be a feeling of irritation around the stoma (the hole created to put the tube into), which could be caused by a build-up of new tissue and blood cells (granulation tissue) growing around site or possibly leakage from the tracheostomy. Your medical team should be able to give you advice on treatment if this occurs.

How long does it take to get used to having a tracheostomy?

It can take time to adapt to living with a tracheotomy especially with communicating and swallowing. Usually, you will initially have the support of a Speech and Language practitioner and a dietician to help with the transition. As you settle with a tracheotomy it is possible to enjoy a good quality of life with a temporary or permanent tracheotomy tube.

How can I talk with a tracheostomy?

Speech can be initially difficult with a tracheotomy. Speech is generated when air passes over the vocal cords at the back of the throat. When a tracheotomy is inserted, most of the air we breathe will pass through the tracheotomy rather than through the vocal cords.

A solution to the problem is wearing a speaking valve on the end of the tracheotomy tube. It is designed to temporarily close every time you breathe out. This allows speech by preventing the air leaking out.

Will I be able to eat normally?

Due to swelling after the initial insertion of the tracheotomy tube, there may be some short-term swallowing difficulties. A speech and language therapist will assess you to help offer advice and teach you techniques to improve your swallowing. Initially, you will be asked to take small sips of fluid, before gradually moving onto puree, soft foods and then your regular diet. Eventually your swallowing ability will improve.

Can I do any physical activity with a tracheostomy?

During the initial 6 weeks after having a tracheotomy procedure, all vigorous physical activity should be avoided. Every day activities should be continued. When you are outdoors, the tracheotomy site should be kept clean, dry and a dressing in place to avoid any risk of infection. A loose piece of clothing such as scarf should be worn to prevent any water or dust entering the tracheostomy. There are surgical bibs which can be supplied from your pharmacy or doctor as needed.

Once you are used to having a tracheostomy it is possible to go back to your usual activities (if your doctor advises it is fine to do so). We have long term tracheostomy patients in the stenosis support group who have adapted equipment to even allow them to go swimming and snorkelling.

How do I cough with a tracheostomy?

Mucus is a natural occurrence in airways, and usually it is dealt with without issue. When there is an interruption, such as the scar tissue in the stenosis or the tracheostomy tube, this can become a problem. If there is an infection or an increase in mucus, there is an increased risk of problems such as a blockage. Ensuring the air breathed in is humidified as much as possible plus regular nebulising can help minimise issues. With practice, you should be able to project mucus through the tube by forcefully breathing out or coughing through the tube.

Patients with tracheotomies quite often cannot cough as well as they need to. A process called suctioning can help keep the airways clear. Suction must be administered by medical professionals unless you have been trained in self-suction.

I'm worried about how I will look with a tracheostomy, and what other people will think. How do other patients deal with this?

It can take time to adapt to having a tracheotomy tube inserted. It is a change in your physical appearance, and you need to allow time to adapt living with a tracheotomy tube.

Many patients initially find it traumatic to adapt to living with a tracheotomy. It is advisable to seek counselling and as well as speaking with family and friends. Your local doctor (and where available, the tracheostomy nurse) can help to refer you for counselling.

People may feel a little uncomfortable seeing someone with a tracheotomy. It is helpful to explain to the reason for having a tracheotomy and how it helps to aid your breathing. Generally, family and friends will adapt very quickly to seeing your tracheotomy.

You can find a more detailed document available in the files section of the support group to help you with looking after your tracheotomy.

Research

Email Catherine if you are interested in taking part in any upcoming research studies related to airway stenosis: airway.stenosis.research@gmail.com

Clinical trials and upcoming studies

- **UK:** Cell therapy that is hoped to have a role in reducing scarring in the airway – engineered tissue that can potentially be grafted into the trachea
- **UK:** An upcoming trial using an oral tablet that may help reduce scarring in the airway
- **Global:** Doctors in Australia are planning to look at the impact of steroid injections on peak flow
- **Global/UK:** Research to improve clinic processes with SGS patients to ensure they are more streamlined, patient led, relevant and address all concerns
- **USA:** Initiated by doctors at Johns Hopkins and Vanderbilt University a blind placebo-drug trial is in progress, being conducted with selected patients during 2022-2023

Appendix

North American Airway Collaborative (NoAAC)

The North American Airway Collaborative is made up of doctors and medical centres around the world (yes, not just North America!) who have an interest in treating subglottic stenosis. They collaborate to conduct research, trials and share information. **These centres and doctors are considered some of the top places to visit for surgery, if you have the choice.**

Doctors from anywhere in the world who are interested in joining NoAAC should make a request to Dr Alexander Gelbard via director@noaac.net

Dr Alexander Gelbard, Dr David Francis, Dr Catherine Gaelyn Garrett, Dr James Netterville, Dr Otis Rickman, Dr Fabian Maldanado	**Vanderbilt University**
Dr Jan Kaserbauer, Dr Eric Edell and Dr Dale Ekbom	**Mayo Clinic (Rochester)**
Dr Robert Lorenz, Dr Paul Bryson and Dr Michael Benninger	**Cleveland Clinic**
Dr Marshall Smith	**University of Utah**
Dr James Damiero	**University of Virginia**
Dr Donald Donovan	**Baylor College of Medicine**
Dr Alexander Hillel	**John Hopkins**
Professor Guri Sandhu	**Imperial College London, UK**
Dr David Lott	**Mayo Clinic (Scottsdale)**
Dr Joshua Schindler	**Oregon Health Sciences University**
Dr Paul Castellanos	Lima, Ohio
Dr Phillip Weissbrod & Dr Samir Makani	University of California, San Diego
Dr Mark Courey	University of California, San Francisco
Dr Matthew Clary & Dr Daniel Fink	University of Colorado
Dr Gregory Postma	Augusta University
Dr Henry Hoffman	University of Iowa
Dr Andrew J McWhorter	Louisiana State University
Dr Ramon Franco	Massachusetts Eye and Ear Infirmary
Dr Robbi Kupfer, Dr Norman Hogikyan and Dr Robert Morrison	University of Michigan
Dr Stephanie N Misono	University of Minnesota

Dr Christopher M Bingcang	University of Nebraska
Dr Rupali N Shah	University of North Carolina
Dr Seth Cohen	Duke University
Dr Ted Mau	University of Texas, SW Dallas
Dr Julina Ongkasuwan, Dr Donald Donovan, Dr Andrew Sikora, Dr Ken Altman	Baylor College of Medicine (Houston)
Dr Albert Merati	University of Washington
Dr David O Francis and Dr Seth H Dailey	University of Wisconsin
Dr David Veivers	University of Sydney, Australia
Dr Brent Richardson	Bastion Voice Institute
Dr Rebecca Howell	University of Cincinnati
Dr Sigriour Sveinsdottir	Landspitali University Hospital, Iceland
Dr Brianna Crawley	Loma Linda Medical Center
Dr Laura Matrka	Ohio State University
Dr Libby Smith	University of Pittsburgh Medical Center
Dr Glenn Shneider	University of Rochester
Dr Ahmed Soliman	Temple University School of Medicine
Dr Joel Blumin & Dr Jonathan Bock	Medical College of Wisconsin
Dr Lindsay Reder and Dr Michael Johns	University of Southern California
Professor Reza Nouraei	Queen’s Medical Centre, Nottingham, UK

Glossary of terms associated with subglottic stenosis

SGS diagnosis and treatment comes with a substantial learning curve and many unfamiliar terms.

Here's an alphabetised list for your reference.

A

AirFlo: AirFlo, an app developed by the North American Airways Collaborative (NoAAC), was a patient airflow monitoring tool (now discontinued) that measured and recorded a patient's peak expiratory flow (PEF) generating data for the Vanderbilt Study.

AmnioFix: This product is an amnion tissue (stem cells) indicated for the treatment of acute and chronic wounds to enhance healing. It is being used by some doctors to try and slow the occurrence of scar tissue. It is still early days for this experimental treatment (around 12 months), so success rates are unknown.

Antinuclear antibody (ANA): ANA is a blood test used to detect the presence of antinuclear antibodies, which may be associated with specific autoimmune diseases such as systemic lupus erythematosus (SLE).

Anti-neutrophil cytoplasmic antibodies (ANCA): ANCA (p-ANCA and c-ANCA) is a blood test for granulomatosis with polyangiitis (GPA, formerly Wegener's granulomatosis).

Awake injections: the common phrase for Awake Serial Intralesional Steroid Injections (SILSI), a procedure pioneered by Dr. Ramon Franco of Massachusetts Eye and Ear Infirmary, where submucosal injections of corticosteroids are made into a stenotic scar with the goal of modifying scars and decreasing operative interventions for SGS treatment (also see Steroid injections). (See The Laryngoscope 2017, Franco et al, "Awake Serial Intralesional Steroid Injections Without Surgery as a Novel Targeted Treatment for Idiopathic Subglottic Stenosis.")

B

Balloon dilation – see Endoscopic tracheal dilation

Bite guard: A bite guard or intubation bite guard is a rigid plastic device inserted in the mouth over the teeth to protect dentition from accidental damage, such as chipping a front tooth, during a surgical procedure. Some SGS patients have had tooth damage and are advised to ensure their surgeon places a bite guard before every airway procedure.

Bronchoscopy: Bronchoscopy is an endoscopic technique of visualizing the inside of the airways for diagnostic and therapeutic purposes using an instrument called a bronchoscopy.

C

Cartilage tracheoplasty: is a form of laryngotrcheoplasty.

Chin stitch: A chin stitch (or Grillo stitch, named for the first surgeon to use this), is a now largely-outdated technique to help keep the head in position after a tracheal resection, only still used at a few centres.

Cilia: Cilia are hair-like structures that line the trachea and help move and expel normal mucus. SGS patients do not have any cilia where the stenotic scar is located, making it more difficult to expectorate.

Clinical trial: see Vanderbilt study

Corticosteroids: Corticosteroids inhibit fibroblast formation and reduce inflammation and can be administered orally by a tablet (e.g. Prednisone), by oral inhalation (e.g. nebulized albuterol, budesonide, i.e. Pulmicort) and by an injection (see Steroid injection and Awake injections). Common corticosteroids used in SGS injection treatment are triamcinolone acetate (Kenalog-40), dexamethasone (Decadron), methylprednisolone (Solu-Medrol) and betamethasone (Celestone).

Cotton-Myer Grading System: This is used to measure the severity of subglottic or tracheal stenosis and is determined with endotracheal tubes demonstrating a leak. It is named after the doctors who first proposed it in 1994.

The grades are:

Grade 0. No or minimal (<10%) discernible stenosis

Grade 1. Discernible obstruction between 10 and 50%

Grade 2. Obstruction between 51 and 70%

Grade 3. Obstruction between 71 and 99%

Grade 4. No discernible lumen

Cricoid cartilage: The cricoid cartilage, or simply cricoid or cricoid ring, is the only complete ring of cartilage around the trachea.

Cricotracheal resection: Cricotracheal resection is a form of open anterior neck surgery. (See: Laryngoscope. 2018 Oct;128(10):2268-2272. doi: 10.1002/lary.27263. Epub 2018 May 14.

'Outcomes after cricotracheal resection for idiopathic subglottic stenosis'. Carpenter PS, Pierce JL, Smith ME.)

C-reactive protein (CRP): CRP is a blood test for inflammation.

Cuff manometer: a device used during general anaesthesia, whether administered using endotracheal intubation or a laryngeal mask airway (LMA), to measure tracheal perfusion pressure (20-30 mmHg). Without a cuff manometer the cuff pressure can increase leading to a situation where tracheal mucosa is no longer perfused, which, in turn, leads to necrosis with subsequent scarring of the tissue and narrowing of the lumen (tracheal stenosis). All patients are advised to request the use of a cuff manometer when undergoing general anaesthesia.

D

Dilation – see Endoscopic tracheal dilation

Direct laryngoscopy: A direct exam of subglottic area via endoscopy (and "in-office scope").

Dyspnea: Shortness of breath, also known as dyspnoea, is a feeling like one cannot breathe well enough with sensations including extra effort/work, chest tightness and/or air hunger.

E

Endoscope: a slender, tubular optical instrument used as a viewing system for examining an inner part of the body and, with an attached instrument, for biopsy or surgery.

Endoscopic balloon dilation – see Endoscopic tracheal dilation

Endoscopic resection – see Endoscopic tracheal resection

Endoscopic tracheal dilation: In cases of subglottic stenosis and tracheal stenosis, endoscopic tracheal dilation or dilatation is the mechanical widening, stretching, or enlarging of the opening of the trachea using a balloon dilator or tracheal dilators (with or without cuts or laser, with or without topical or injected corticosteroids, with or without Mitomycin-C).

Endoscopic tracheal reconstruction: Named for Jan Maddern (the first patient to undertake this procedure), "the Maddern" is an endoscopic transoral surgical procedure where the scar is removed, and a skin graft is placed temporarily into the trachea for two weeks to encourage healthy cells to regrow in place of the diseased tissue.

Endoscopic tracheal resection: A multidisciplinary team at Mayo Clinic in Rochester, USA developed this technique which involves transoral resection of the scar tissue using a laser. Longitudinal research by Vanderbilt University and the NoAAC has found this resection technique to deliver long lasting results for patients without compromising voice. It is slowly being adopted by other centres globally. (This technique is described in detail in: Maldonado F, et al. Idiopathic subglottic stenosis: An evolving therapeutic algorithm. Laryngoscope. 2014;124:498.)

Endoscopy: An endoscopy is a procedure to look inside the body using an endoscope to examine the interior of a hollow organ or cavity of the body.

Endotracheal intubation: Endotracheal or tracheal intubation, usually simply referred to as intubation, is the placement of an endotracheal tube into the trachea to maintain an open airway or to serve as a conduit through which to administer certain drugs.

Endotracheal tube (ETT): A flexible plastic tube placed into the trachea during endotracheal intubation to establish and maintain airway patency, prevent aspiration into the lungs, i.e. to "secure" the airway and allow for mechanical ventilation. ETT sizes (e.g., 5.5, 6.0, 6.5, etc.) refer to the inner diameter of the tube measured in mm and vary by and within gender. Standard practice is to use a 7.0 ETT for women, but it may be too large for some airways, and improper ETT sizing is known to cause subglottic stenosis. SGS patients may wish to add their recommended ETT size to a Medical Alert ID.

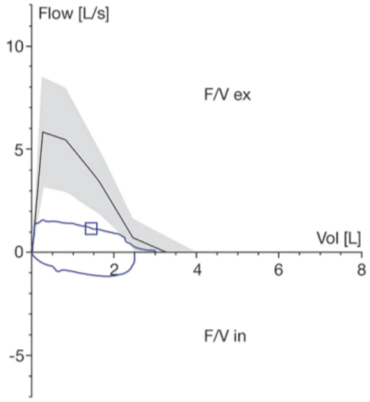
Erythrocyte sedimentation rate (ESR): ESR is a blood test for inflammation.

F

Fibroblast: a cell in connective tissue which produces collagen and other fibres.

Flow Volume Loop: A flow volume loop is a visual representation of spirometry data that shows whether airflow is appropriate for a particular lung volume. SGS patients have "flattened" flow volume loops.

(Right: A typical flow volume loop of a patient with subglottic stenosis – source: Mayo Clinic)



Flutter valve: Flutter valve is an airway clearance device to help expel mucus by combining positive expiratory pressure (PEP) therapy with airway vibrations.

G

Glottis: The glottis is the opening between the vocal folds.

Granulomatosis with polyangiitis (GPA): GPA, formerly Wegener's Granulomatosis, is an auto-immune collagen vascular disease.

H

Huff coughing: an alternative to deep coughing if you have trouble clearing your mucus. See section on reducing inflammation and mucus

I

Idiopathic subglottic stenosis (SGS or ISS): refers to narrowing of the upper trachea of unknown cause. The disease is rare, with an estimated incidence of 1 per 400,000 person-years. SGS accounts for approximately 19 percent of patients with laryngotracheal stenosis, with the majority having an identifiable cause such as an iatrogenic injury, neck or throat trauma, or an inflammatory connective tissue disease. The exact pathogenesis of ISS is unknown.

IL-23/IL-17A Axis: The relationship between Interleukin-23 (IL-23) and Interleukin 17A (IL-17A), small proteins called cytokines, is referred to as the IL-23/IL-17A axis. The aberrant mucosal immune activation seen in the large airway of SGS patients helps us understand the molecular pathogenesis of SGS and may lead to more directed targeting with drugs (see The Laryngoscope 2016, Gelbard et al, "Idiopathic Subglottic Stenosis is Associated with Activation of the Inflammatory IL-17A/IL-23 Axis").

Indirect laryngoscopy: uses a small mirror held at the back of your throat. The health care provider shines a light on the mirror to view the throat area. This is a simple procedure. Most of the time, it can be done in the provider's office while you are awake. A medicine to numb the back of your throat may be used.

In-office injections – see Awake injections

Intubation – see Endotracheal intubation

J

Jet ventilation: Used in conjunction with general anaesthesia, jet ventilation is a transoral means of ventilation during laryngeal surgery and procedures on the upper airway.

K

Kenalog – see Corticosteroids

L

Laryngeal mask airway (LMA): also known as laryngeal mask— is a medical device that keeps a patient’s airway open during anaesthesia or unconsciousness and can be used as an alternative to intubation during non-airway surgery.

Laryngoscope: A laryngoscope is a rigid or flexible endoscope passed through the mouth and equipped with a source of light and magnification, for examining and performing local diagnostic and surgical procedures on the larynx.

Laryngoscopy: Laryngoscopy is endoscopy of the larynx, a part of the throat, used to view the vocal folds and glottis.

Laryngotracheal reconstruction: Laryngotracheal (luh-ring-go-TRAY-key-ul) reconstruction surgery widens your windpipe (trachea) to make breathing easier. Laryngotracheal reconstruction involves inserting a small piece of cartilage — stiff connective tissue found in many areas of your body — into the narrowed section of the windpipe to make it wider.

Laryngotracheal stenosis (LTS): Laryngotracheal stenosis, a fixed extrathoracic obstruction, is the abnormal narrowing of the central air passageways, which can occur at the level of the larynx, trachea, carina or main bronchi. SGS patients may also have stenosis at the glottis and/or supraglottic stenosis.

Laryngotracheoplasty: a form of open anterior neck surgery that includes both cartilage tracheoplasty and slide tracheoplasty. It is also known as laryngotracheal reconstruction.

Larynx: The larynx or voice box is an organ in the top of the neck involved in breathing, producing sound, and protecting the trachea against food aspiration. It houses the vocal folds, and manipulates pitch and volume, which are essential for phonation.

Living with Idiopathic Subglottic Stenosis: “Living with Idiopathic Subglottic Stenosis” (www.Facebook.com/groups/IdiopathicSubglotticStenosis) is a Facebook support group for patients and their families started in 2009 by Catherine Anderson, the NoAAC Patient Partner.

M

Maddern – see Endoscopic tracheal reconstruction

Mayo protocol: The Mayo protocol is an SGS treatment approach that combines antibiotics to suppress inflammation (e.g., Bactrim, azithromycin or erythromycin), corticosteroids to inhibit fibroblast formation and reduce inflammation, and proton-pump inhibitors (PPIs) to reduce gastric acid reflux (gastroesophageal reflux disease or GERD).

Mitomycin-C: Mitomycin-C is a chemotherapeutic agent that can be applied to a stenotic scar at the time of an endoscopic tracheal dilation, although there is no scientific evidence of its effectiveness.

Mucus plug: Mucus impaction or plugging occurs when mucus becomes thick and sticky creating a mass that is difficult to expectorate blocking the airway and possibly resulting in death. Mucus plugs can be avoided by respiratory therapy, medication, and mucolytic therapy.

Mucolytic therapy: helps expel mucus, can include a combination of mucolytic medication (Over the counter (OTC) expectorant cough syrup, throat lozenge with glycerine, pectin, or slippery elm not menthol or eucalyptus; amino acid N-Acetyl Cysteine (NAC) or NAC as a capsule or nebulized NAC, e.g. Mucomyst) and the use of an airway clearance technique (‘huff’ cough ‘huffing’ exercise) or device (flutter valve).

Mycobacterium tuberculosis (MtbC): MtbC is a pathogenic bacteria in the family Mycobacteriaceae and the causative agent of tuberculosis, yet rather than MtbC, a variant member of the MtbC complex or a closely related novel mycobacterium has been found to be present in SGS tracheal biopsy specimens (see The Laryngoscope 2016, Gelbard et al, “Molecular Analysis of Idiopathic Subglottic Stenosis for Mycobacterium Species”).

N

Nebulizer: A nebulizer is a saline and/or drug delivery device used to administer medication (or saline) in the form of a mist inhaled into the airway to thin mucus making it easier to expectorate.

North American Airway Collaborative (NoAAC): NoAAC (noaac.net) is a collective of medical centres who are collaborating to investigate the underlying cause of SGS and seeking to provide patients with information that will improve their quality of life through informed decision making. Its leadership team of multi-disciplinary clinicians, surgeons, patient representatives, and health care research personnel volunteers its time and expertise. NoAAC was founded at Vanderbilt University and its director is Dr. Alexander Gelbard. The NoAAC Patient Partner Catherine Anderson established “Living with Idiopathic Subglottic Stenosis” and the NoAAC Patient Representative Kesi-Dorner Wright established the Tracheal Stenosis Foundation.

O

Open anterior neck surgery: includes four surgical procedures: cricotracheal resection, Laryngotracheoplasty (cartilage tracheoplasty and slide tracheoplasty), REACHER and tracheostomy.

Otolaryngology: Otolaryngology is a surgical subspecialty that deals with the ear, nose, and throat (ENT) and related structures of the head and neck. Doctors who specialize in this area are called otolaryngologists, ENT doctors, ENT surgeons or head and neck surgeons.

P

Patient Centered Outcomes Research Institute (PCORI): PCORI (pcori.org) funds research that can help patients and those who care for them make better-informed decisions about the healthcare choices they face every day, guided by those who will use that information. PCORI funded the Vanderbilt Study.

Peak expiratory flow (PEF): PEF is a person's maximum speed of expiration measured with a peak flow meter.

Peak flow meter: A peak flow meter is a portable, hand-held device that measures how well your lungs expel air. It measures your ability to push air out of your lungs in one fast blast by blowing hard through a mouthpiece (i.e. 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).

Phonation: Phonation is the process by which the vocal folds produce certain sounds through periodic vibration.

Pulmonary Function Test (PFT): PFT, also called spirometry or lung function testing, is a group of tests that evaluate the respiratory system, including the function of lung capacity, chest wall mechanics, etc.

Pulse oximeter: Pulse oximeter is a device placed on your finger to measure oxygen saturation.

R

REACHER (Retrograde, endoscopically assisted cricoid hypertrophic epithelial resection): REACHER is an open anterior neck surgical procedure pioneered by Dr Robert Lorenz of Cleveland Clinic, Ohio USA, where the diseased lining of the cricoid is removed and the outer cartilage is preserved.

Rough Guide: Idiopathic Subglottic Stenosis – A Rough Guide for Beginners by Catherine Anderson, the NoAAC Patient Partner, is a reference manual for SGS patients and their doctors.

S

Saline: also known as saline solution, is a mixture of sodium chloride in water (vials come in 0.9% and 3.0% strengths) that can be administered orally with a nebulizer.

Slide tracheoplasty: a type of laryngotracheoplasty, is a surgical procedure used for more severe grades of long-segment tracheal stenosis or complete tracheal rings where the narrowed trachea is divided across the middle of the stenosis. The back of the lower tracheal segment and front of the upper tracheal segment are cut, and the opened ends are then slid onto each other and sutured in place to create an airway that is twice as wide and half as long.

SLT/SLP: see speech and language therapist/pathologist

Speech and Language Therapist/Pathologist: Provide treatment, support and care for people who have difficulties with communication, eating, drinking and swallowing.

Spirometry: Spirometry, the most common pulmonary function test, measures lung function, specifically the amount and/or speed of air that can be inhaled and exhaled.

Steam inhalation: Steam inhalation is a method of introducing warm, moist air into the lungs via the nose and throat for therapeutic benefit such as thinning mucus by breathing into a portable device. Essential oils can also be used.

Stenosis: an abnormal narrowing or contraction of a body passage or opening.

Steroid injection: refers to the submucosal injection of corticosteroids into the stenotic scar administered either in an office setting (externally through the neck or via transnasal endoscope) or in an operative setting with or without dilation (under moderate sedation or under general anaesthesia via transoral endoscope or transnasal videoscope).

Stoma: any opening in the body. A patient with a tracheotomy has a stoma in their trachea.

Stridor: Stridor is a high-pitched breath sound (commonly called the "Darth Vader" sound) resulting from turbulent air flow in the larynx or lower in the bronchial tree, caused by a narrowed or obstructed airway.

Subglottic stenosis: Narrowing of the subglottis is known as subglottic stenosis.

Subglottis: the subglottis or subglottic region is the lower portion of the larynx, extending from just beneath the vocal folds down to the top of the trachea.

Supraglottic stenosis: Supraglottic stenosis is narrowing of the airway above the glottis,

T

Trachea: or windpipe is a cartilaginous tube that connects the pharynx and larynx to the lungs, allowing the passage of air, and branches into the two primary bronchi.

Tracheal reconstruction: a surgical procedure to remove a portion of the trachea with a long stenotic scar and to insert a stent or piece of rib into the neck to replace the lost cartilage and prevent the neck from collapsing.

Tracheal resection: a surgical procedure to remove a portion of the narrowed portion of a patient's trachea or windpipe through an incision in the neck and reattaching the upper and lower healthy parts with stitches.

Tracheal stenosis – see Laryngotracheal stenosis

Tracheal T-tube: a silicone stent for the trachea with an external limb.

Tracheal Stenosis Foundation (TSF): TSF is a non-profit organization founded by patient Kesi Dorner-Wright that raises awareness and holds an (almost) annual patient-centered conference in the USA whose speakers include members of the NoAAC leadership team.

Tracheotomy: also known as tracheostomy is a surgical procedure that consists of making an incision on the anterior aspect of the neck and opening a direct airway through an incision in the trachea.

V

Vanderbilt Study: The commonly called Vanderbilt Study is an ongoing clinical trial (2015-2018) formally known as the **'**NoAAC PR-02 SGS Clinical Trial - Treatment Alternatives in Adult Rare Disease; Assessment of Options in Idiopathic Subglottic Stenosis' (https://clinicaltrials.gov/ct2/show/NCT02481817). Dr. Alexander Gelbard of Vanderbilt University is the study's Principal Investigator.

Videoscope: A videoscope, also called a transnasal endoscope or distal chip video esphagoscope, is a medical device that records video of a procedure, such as transnasal steroid injections.

Vocal folds: Vocal folds or vocal cords are composed of twin folds of mucous membrane stretched horizontally, from back to front, across the larynx.

W

Wegener's Granulomatosis (WG) - see Granulomatosis with polyangiitis (GPA)

Reading List

Being diagnosed and living with a rare, life-threatening disease can be overwhelming at times. Educating oneself about the disease, past and current research, and evolving treatment options helps to make an informed patient. With knowledge, you can ask your airway surgeon questions and make informed decisions about your treatment.

The following is a list of SGS-related articles published in academic journals. (The articles appear in reverse chronological order with the most recent publications first.) The list is compiled from various sources including posts on the "Living with Idiopathic Subglottic Stenosis" support community and documents in its "Files" tab. This is not intended to be a full or complete list of all published articles on SGS, but it is a selection of many relevant ones. Each listing includes the title, its authors, publication details (journal name, volume, page numbers, and date of publication), and its Digital Object Identifier (DOI). The DOI alphanumeric string identifies an academic article and provides a persistent link to its location on the Internet. You can go to the DOI website (https://doi.org) and enter any DOI to retrieve an article.

We have included the authors' names so you can familiarize yourself with the leading airway surgeons and researchers in this field, many of whom are participating members of the North American Airway Collaborative. For those of you who have attended any of the Tracheal Stenosis Foundation conferences or the 2018 presentation of the SGS clinical trial results, you may have met some of these individuals in person. You may also recognize many of the authors' names from social media posts as the surgeons who care for many of us. These devoted and talented surgeons also conduct SGS research and publish their findings in peer-reviewed journals. We all hope their collective work leads to better understanding of SGS and eventually a cure.

The articles are technical in nature and you may find yourself wondering how to understand it all. If you read the articles' abstracts and conclusions and then work your way into the science and all the details, you should be able to grasp the overall concepts. Many articles appear in *The Laryngoscope*, which may just become your favourite new reading material.

The Articles

2022

"The Effect of Subglottic Steroid injection on Blood Glucose in a Cohort of Patients With Subglottic Stenosis"
Bensoussan Y, Martinez E, Van Der Woerd B, Johns M, Nguyen C, Watts S, O'Dell K.
Laryngoscope. Sep 27 2022.
DOI: 10.1002/lary.30392.

"'Is there something wrong with your voice?' A qualitative study of the voice concerns of people with laryngotracheal stenosis"
Gemma M Clunie, Athina Belsi, Justin W. G Roe, Guri Sandhu, Alison McGregor, Caroline M. Alexander
International Journal of Language and Communication Disorders, September 2022
DOI: 10.1111/1460-6984.12794

""A Major Quality of Life Issue": A Survey-Based Analysis of the Experiences of Adults with Laryngotracheal Stenosis with Mucus and Cough"
Gemma Clunie; Catherine Anderson; Justin W.G. Rose; Guri Sandhu; Alison MacGregor; Caroline M. Alexander
Annals of Otology, Rhinology & Laryngology September 2022
DOI: 10.1177/00034894211050627

"New Treatment Strategy for Subglottic Stenosis Using the Trachealator, a Novel Non-occlusive Balloon"
Leonie G. M. Wijermars, MD, PhD ; Carlijn E. L. Hoekstra, MD, PhD; Thao T. T. Nguyen, MD; Markus F. Stevens, MD, PhD; Frederik G. Dikkers, MD, PhD
The Laryngoscope, May 2022
DOI: 10.1002/lary.30234

"An Updated Review of Subglottic Stenosis: Etiology, Evaluation,and Management"
Luke J. Pasick, Mursalin M. Anis, David E. Rosow
Interventional Pulmonology, February 2022
https://doi.org/10.1007/s13665-022-00286-6

"Persistent Throat Symptoms Should Not Be Treated With Pills That Reduce Stomach Acid"
James O'Hara
Ear, Nose and Throat, January 13, 2022
DOI: 10.3310/alert_48810

"Dilation Versus Laser Resection In Subglottic Stenosis: Protocol For A Prospective International Multicentre Randomised Controlled Trial (AERATE Trial)"
Thibaud Soumagne; Nicolas Guibert; Ihab Atallah; Yves Lacasse; Hervé Dutau; Marc Fortin
BMJ Open, 12:e053730, 2022
DOI: 10.1136/bmjopen-2021-053730

"Laryngotracheal Stenosis: Mechanistic Review"
Delaney J. Carpenter, MD; Osama A. Hamdi, MD; Ariel M. Finberg, BS; James J. Daniero, MD, MS
Head & Neck, 1–13, Basic Science Review, 2022
DOI: 10.1002/hed.27079

"Idiopathic Subglottic Stenosis Arises At The Interface Of Host And Pathogen"
Alexander Gelbard; Meghan H. Shilts; Britton Strickland; Kevin Motz; Hsiu-Wen Tsai; Helen Boone; Wonder P. Drake; Celestine Wanjalla; Paula Marincola Smith; Hunter Brown; Marisol Ramierez; James B. Atkinson; Jason Powell; A. John Simpson; Seesandra V. Rajagopala; Simon Mallal; Quanhu Sheng; Alexander T. Hillel; Suman R. Das
BMJ, February 4, 2022
DOI: 10.1101/2022.02.02.22270308

"How A Potent Antifibrotic Peptide Works And Why It Could Reverse Scarring In Multiple Organs"
Caren Doueiry
Medical University of South Carolina, January 10, 2022
https://medicalxpress.com/news/2022-01-potent-antifibrotic-peptide-reverse-scarring.html

2021

“The Liverpool Experience: The Role of Immunosuppression in treating Vasculitic Subglottic Stenosis”
Matthew Zammit, Vedika Dhunnoo, Andrew Kinshuck, Sarah Hardy, Janice Harper, Alessandroo Panarese, and Christopher Webb
Clinical Otolaryngology, September 2021
DOI: 10.1111/coa.13900

“Supraglottic Stenosis: What Do We Know About Swallow Function in This Population?”
Avisha Daryanini; Gemma Clunie; Lindsay Lovell; Gupreet Sandhu; Chadwan Al-Yagchi, Justine Roe
Imperial College Healthcare, 2021

“Antifibrotic Role of Nintedanib In Tracheal Stenosis After a Tracheal Wound”
Yuhua Fan, MD; Xin Li, MD; Xing Fang, MD; Yalan Liu, PhD; Suping Zhao, MD; Zicheng Yu, PhD; Yaoyun Tang, MD; Ping Wu, MD
The Laryngoscope, 131:9, E2496-E2505, September 2021
DOI: 10.1002/lary.29618

“Prevalence And Incidence Of Idiopathic Subglottic Stenosis In Southern And Central Alberta: A Retrospective Cohort Study”
Ryan K. Chan; Beau Ahrens; Paul MacEachern; J. Douglas Bosch; Derrick R. Randall
Journal of Otolaryngology - Head & Neck Surgery, 50:64, 2021
DOI: 10.1186/s40463-021-00544-8

“Monitoring Adult Subglottic Stenosis With Spirometry And Dyspnea Index: A Novel Approach”
Eleftherios Ntouniadakis, MD; Josefin Sundh, MD, PhD; Mathias von Beckerath, MD, PhD
Otolaryngology– Head and Neck Surgery, 1–7, 2021
DOI: 10.1177/01945998211060817

“Serial Intralesional Steroid Injection For Subglottic Stenosis: Systemic Side Effects And Impact On Surgery-Free Interval”
Andrew J. Neevel; Ari D. Schuman, MD; Robert J. Morrison, MD; Norman D. Hogikyan, MD; Robbi A. Kupfer, MD
OTO Open, Vol. 5(4) 1–8, 2021
DOI: 10.1177/2473974X211054842

“A Major Quality Of Life Issue”: A Survey-Based Analysis Of The Experiences Of Adults With Laryngotracheal Stenosis With Mucus And Cough”
Gemma M. Clunie, MSc; Catherine Anderson, BA, PGD; Matthew Savage, BSc; Catherine Hughes, BSc; Justin W.G. Roe, PhD, FRCSLT; Gurpreet Sandhu, MD; Alison McGregor, PhD; Caroline M. Alexander, PhD, MSc, MCSP
Annals of Otology, Rhinology & Laryngology, 1–9, 2021
DOI: 10.1177/0003489421105062

“Laryngotracheal Resection Can Be Performed Safely Without A Guardian Chin Stitch—A Single-Centre Experience Including 165 Consecutive Patients”

Thomas Schweiger; Matthias Evermann; Imme Roesner; Anna-Elisabeth Frick; Doris-Maria Denk-Linnert; Walter Klepetko; Konrad Hoetzenecker
European Journal of Cardio-Thoracic Surgery, 00:1–7, 2021
DOI: 10.1093/ejcts/ezab092

"Not Just Dyspnoea: Swallowing As A Concern For Adults With Laryngotracheal Stenosis Undergoing Airway Reconstruction"
Gemma M. Clunie, Athina Belsi, Justin W. G. Roe, Caroline M. Alexander, Gurpreet Sandhu, Alison McGregor
Dysphagia 2021
DOI: 10.1007/s00455-021-10287-3

"Impact of Adjuvant Medical Therapies on Surgical Outcomes in Idiopathic Subglottic Stenosis"
Matthew R. Hoffman, MD, PhD; Ankita Patro, MD, MS; Li-Ching Huang, PhD; Sheau-Chiann Chen, PhD; Lynn D. Berry, PhD; Alexander Gelbard, MD; David O. Francis, MD, MS
The Laryngoscope, 00:1–7, 2021
DOI: 10.1002/lary.29675

"Down-Regulation of a Profibrotic Transforming Growth Factor-b1/Cellular Communication Network Factor 2/Matrix Metalloprotease 9 Axis by Triamcinolone Improves Idiopathic Subglottic Stenosis"
J. Humberto Treviño-Villarreal, Justin S. Reynolds, P. Kent Langston, Andrew Thompson, James R. Mitchell, Ramon A. Franco, Jr.
The American Journal of Pathology, 191:8, August 2021
DOI: 10.1016/j.ajpath.2021.05.013

"Endoscopic Resection and Mucosal Reconstitution With Epidermal Grafting: A Pilot Study in Idiopathic Subglottic Stenosis"
Ruth J. Davis, MD; Ioan Lina, MD; Kevin Motz, MD; Alexander Gelbard, MD; Robert R. Lorenz, MD; Guri S. Sandhu, MD; Alexander T. Hillel, MD
Otolaryngology– Head and Neck Surgery, 1–10, 2021
DOI: 10.1177/01945998211028163

2020

"A Look at the Latest Research on Subglottic Stenosis"
Nikki Kean
ENT Today, August 17, 2020
https://www.enttoday.org/article/a-look-at-the-latest-research-on-subglottic-stenosis/

"Endoscopic Wedge Excisions With CO2 Laser For Subglottic Stenosis"
Dale C. Ekbom, MD; Semirra L. Bayan, MD; Andrew J. Goates, MD; Jan L. Kasperbauer, MD
The Laryngoscope, 00:1-5, 2020
DOI: 10.1002/lary.29013

"Impact of the COVID-19 Pandemic on Patients with Idiopathic Subglottic Stenosis"
Anderson C, Sandhu G, Yaghchi CA..
Ear, Nose & Throat Journal. December 2020.
DOI:10.1177/0145561320977467

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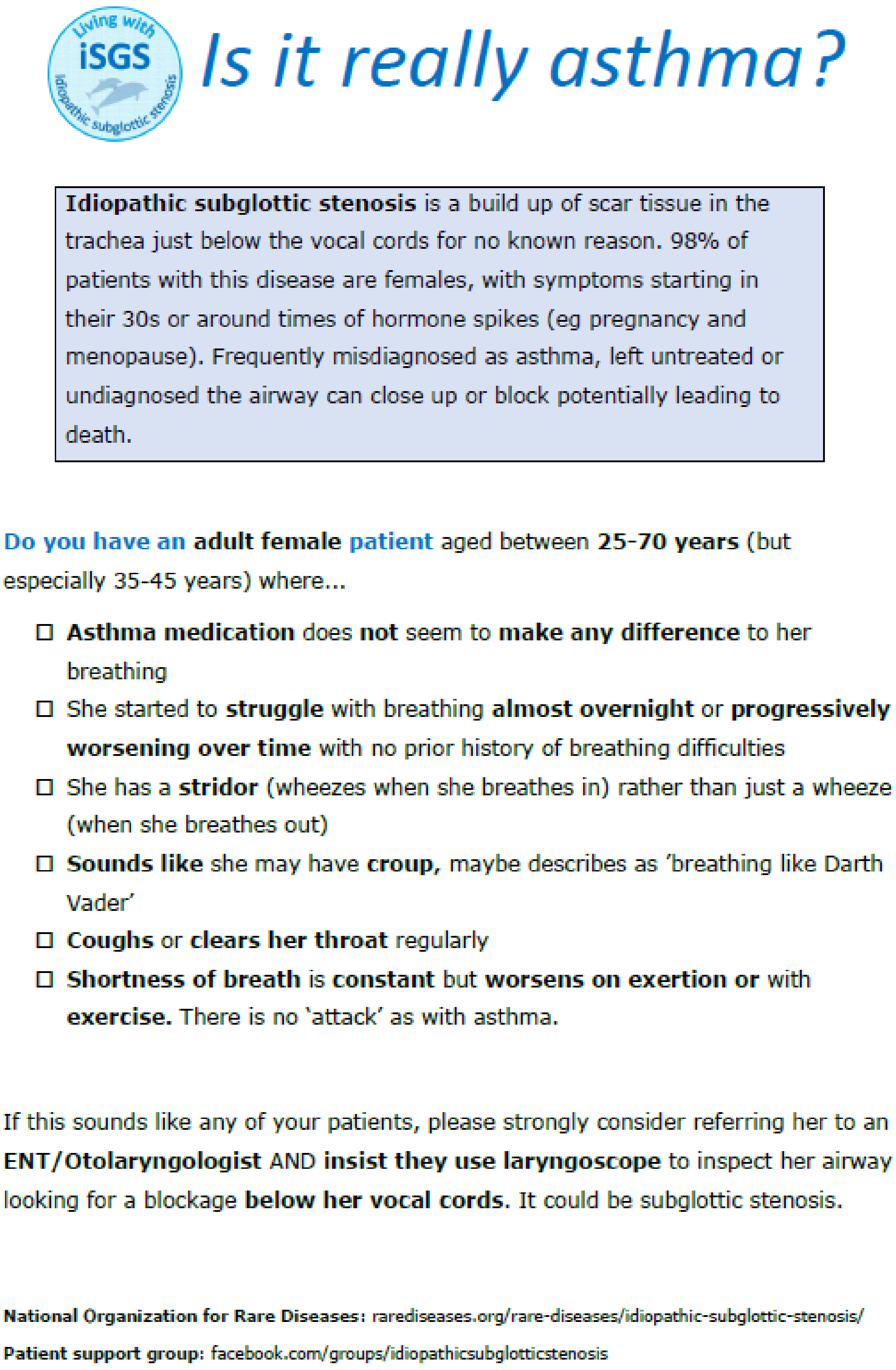
Resources

Please share this document with your primary care doctor/GP



Is it really asthma?

Idiopathic subglottic stenosis is a build up of scar tissue in the trachea just below the vocal cords for no known reason. 98% of patients with this disease are females, with symptoms starting in their 30s or around times of hormone spikes (eg pregnancy and menopause). Frequently misdiagnosed as asthma, left untreated or undiagnosed the airway can close up or block potentially leading to death.

Do you have an adult female patient aged between **25-70 years** (but especially 35-45 years) where...

- ☐ **Asthma medication** does **not** seem to **make any difference** to her breathing
- ☐ She started to **struggle** with breathing **almost overnight** or **progressively worsening over time** with no prior history of breathing difficulties
- ☐ She has a **stridor** (wheezes when she breathes in) rather than just a wheeze (when she breathes out)
- ☐ **Sounds like** she may have **croup,** maybe describes as 'breathing like Darth Vader'
- ☐ **Coughs** or **clears her throat** regularly
- ☐ **Shortness of breath** is **constant** but **worsens on exertion or** with **exercise.** There is no 'attack' as with asthma.

If this sounds like any of your patients, please strongly consider referring her to an **ENT/Otolaryngologist** AND **insist they use laryngoscope** to inspect her airway looking for a blockage **below her vocal cords.** It could be subglottic stenosis.

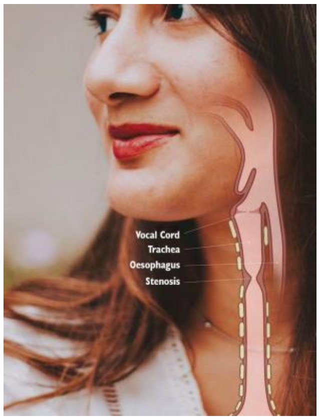
National Organization for Rare Diseases: rarediseases.org/rare-diseases/idiopathic-subglottic-stenosis/

Patient support group: facebook.com/groups/idiopathicsubglotticstenosis

Dear Doctor, I have subglottic stenosis….

The following page is designed to help provide an explanation of your airway disease for a doctor who may not be familiar with treating someone with subglottic stenosis – such as a new general practitioner or perhaps a doctor in an emergency room (even if they are an ear, nose and throat specialist). You can complete the relevant details and bring it with you to explain your condition when you might see a clinician other than your usual specialist.

Dear Doctor



This patient has a rare airway disease, **idiopathic subglottic stenosis**. They may be presenting to you today because they have **issues with breathing** and are unable to see their usual treating otolaryngologist consultant immediately.

Idiopathic subglottic stenosis (SGS) is a rare disease consisting of upper airway obstruction which nearly universally occurs in adult, Caucasian women. Symptoms vary from dyspnoea, stridor, dysphonia and cough. There is no known cause for this, though it could be related to autoimmune disease (e.g. GPA), hormonal sensitivity or previous intubation if there is history.

The stenosis (scar tissue) is located just below the vocal cords, but it is possible for it to spread to other areas of the trachea, including within or above the vocal cords, or lower down in the trachea. A stenosis which is often quite stable, needing only annual intervention can suddenly become inflamed, or more aggressive and close rapidly without apparent cause. It is important to investigate before sending this patient away.

Typical treatments include a balloon dilation and steroid injections into the affected area.

Please do not consider a tracheotomy without prior discussion with the patient's consulting surgeon unless it is deemed potentially lifesaving by the local airway (otolaryngology/anaesthetic/ER consultant) team i.e. an emergency. This could potentially cause further damage to the airway and worsening stenosis longer term.

There are potentially several different reasons for their current breathing issues, which could be assessed:

Issue	Explanation
Non-COVID 19 Infection	Given the area of narrowing in her trachea, there is a high risk of upper airway infection. This increases risk of a life-threatening mucus plug if left untreated.
COVID-19 Infection	SGS patients may be more at risk of complications from coronavirus infection due to impaired mucociliary clearance and unknown immune abnormalities. Breathing problems should not be assumed to be due to the stenosis, and tests to rule out COVID 19 infection carried out. Management of SGS patients with coronavirus infection should follow local and national guidelines for

	patients without such stenosis. Consideration of 2-3 week delay before deciding to perform elective tracheostomy should be made, in accordance with guidance, in the unlikely event of ventilation being required.
Mucus plug	Mucus builds up behind the scar tissue and can become thickened. If left untreated this can block the stenosis, restricting airflow and potentially leading to asphyxiation. Assess immediate risk of airway compromise and consider urgent referral to an otolaryngologist/ ENT surgeon for bronchoscopy and/or removal of plug. Frequent saline nebulising potentially with an expectorant (e.g. Guaifenesin, Robitussin or Mucinex) designed to break down mucus. Most airway stenosis patients have a saline nebuliser at home which could be used regularly in this situation.
Inflammation	Inflammation can be triggered by two factors: 1.Due to the potential autoimmune impact of this disease, airway inflammation is common in SGS patients and can occur for no obvious reason. This can further constrict an already narrow airway. 2. SGS patients can be at increased risk of laryngeal reflux which can lead to inflammation in the throat.
Closure of stenosis	In some patients, the scar can close rapidly (often sometimes within weeks of an otherwise successful dilatation surgery). The signs of this are a rapid and consistent fall in PEF rate, significant stridor, and respiratory distress. Assess immediate risk of airway compromise and urgently refer to an otolaryngologist/ ENT surgeon for management. Alert the consultant anaesthetist on-call.
New stenosis	Occasionally patients will develop a second stenosis. The patient may notice a change in their breathing which is perhaps different to before. This can be most accurately diagnosed with a bronchoscopy or a fine slice CT of the neck.

Key details about this patient:

Name					
Age/DOB		**Height**		**Weight**	
Expected Peak Expiratory Flow (PEF) for age/height/ gender if healthy (a)		**Current PEF (b)**			

Name of usual treating otolaryngologist		**Contact details**	
Current medications			
Additional conditions			

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