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Idiopathic Subglottic Stenosis

A Rough Guide for Beginners



Last updated: May 2020

Please join the Facebook Support group:

www.Facebook.com/groups/IdiopathicSubglotticStenosis

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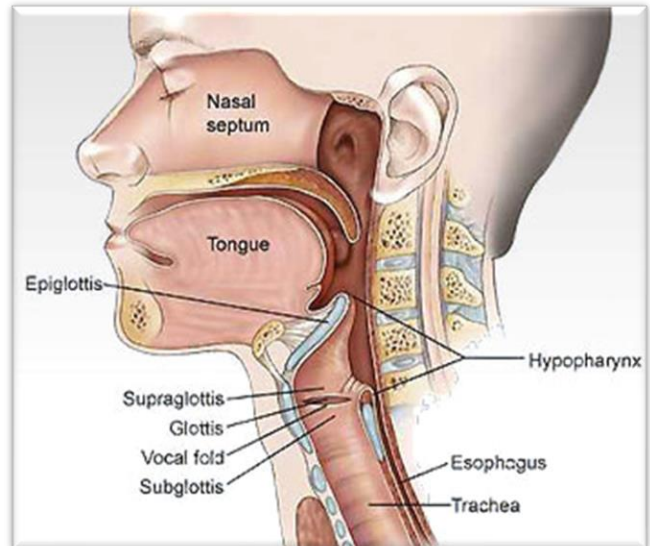
What is idiopathic subglottic stenosis?

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

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

Getting down to basics;

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



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

This document will help you regardless of your stenosis 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 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
- Thyroid or major anterior neck surgery
- Exposure to radiation on your neck
- Caustic (eg chemical burn) or thermal (heat) injuries to the airway
- Vasculitis (a condition that involves inflammation of the blood vessels)
- Angiotensin converting enzyme (ACE) and antinuclear cytoplasmic antibody (ANCA)



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



Polytrauma - Patients 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.

What are the common symptoms of iSGS?

We all share some common symptoms:

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

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

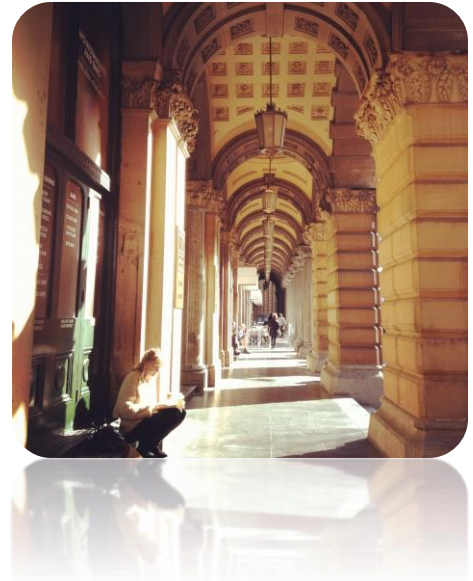
Often we as sufferers stop noticing this sound ourselves, and it is friends, relatives and work colleagues who might mention it to us. We often describe this as Darth Vader breathing!

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

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

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

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



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 report temporary feelings of euphoria once they can breathe well.

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

Who gets iSGS?

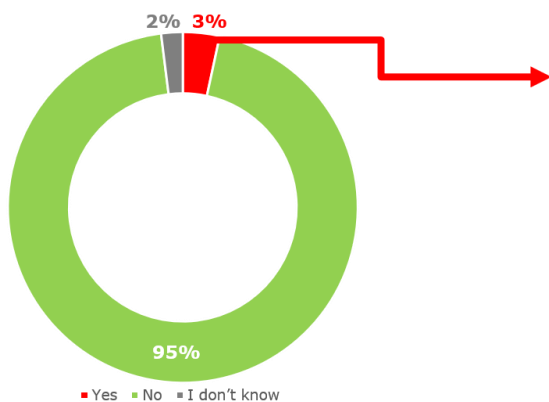
Idiopathic subglottic stenosis is very rare – less than one in 200,000 people get this disease (the definition of 'rare') – it is in fact estimated to only impact one person in half a million per year.

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.

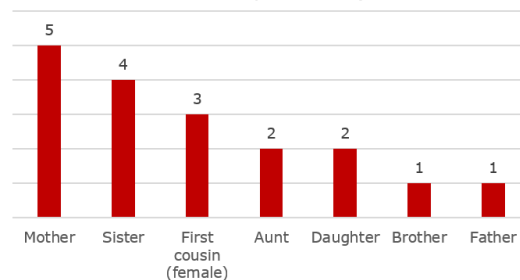
People are often afraid of passing this on to their children – that is very unlikely. Just 3% of iSGS patients have a blood relative with this disease, suggesting this is not something you should be too worried about. If a relative (particularly a female) has similar symptoms to you, then they should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

Blood relatives with iSGS



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

Blood relative with iSGS (Caution: small sample n=17)



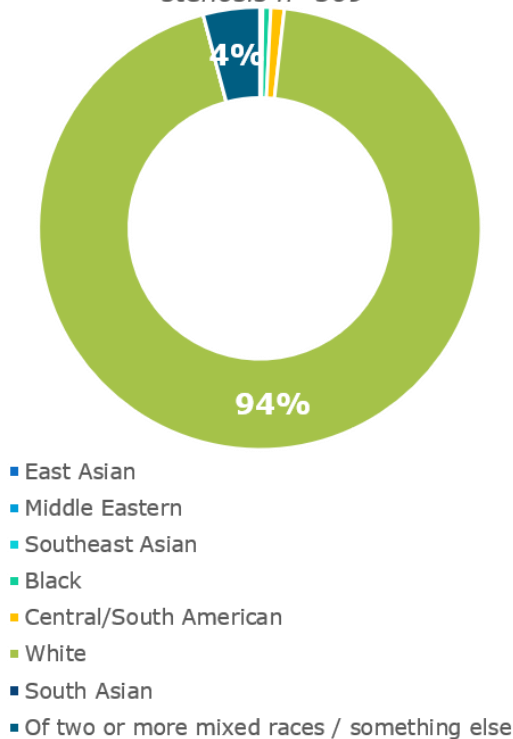
Q31. What type of subglottic stenosis were they diagnosed with? Base: Patients with idiopathic subglottic stenosis who indicated they had a blood relative also with a stenosis n=17

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:

How would you describe your ethnic background?

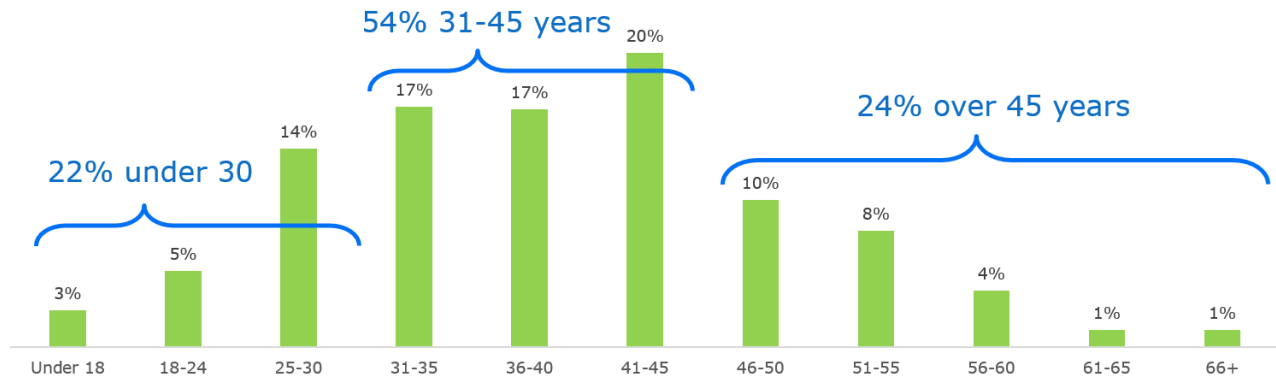
Base: Patients with idiopathic subglottic stenosis n=509



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

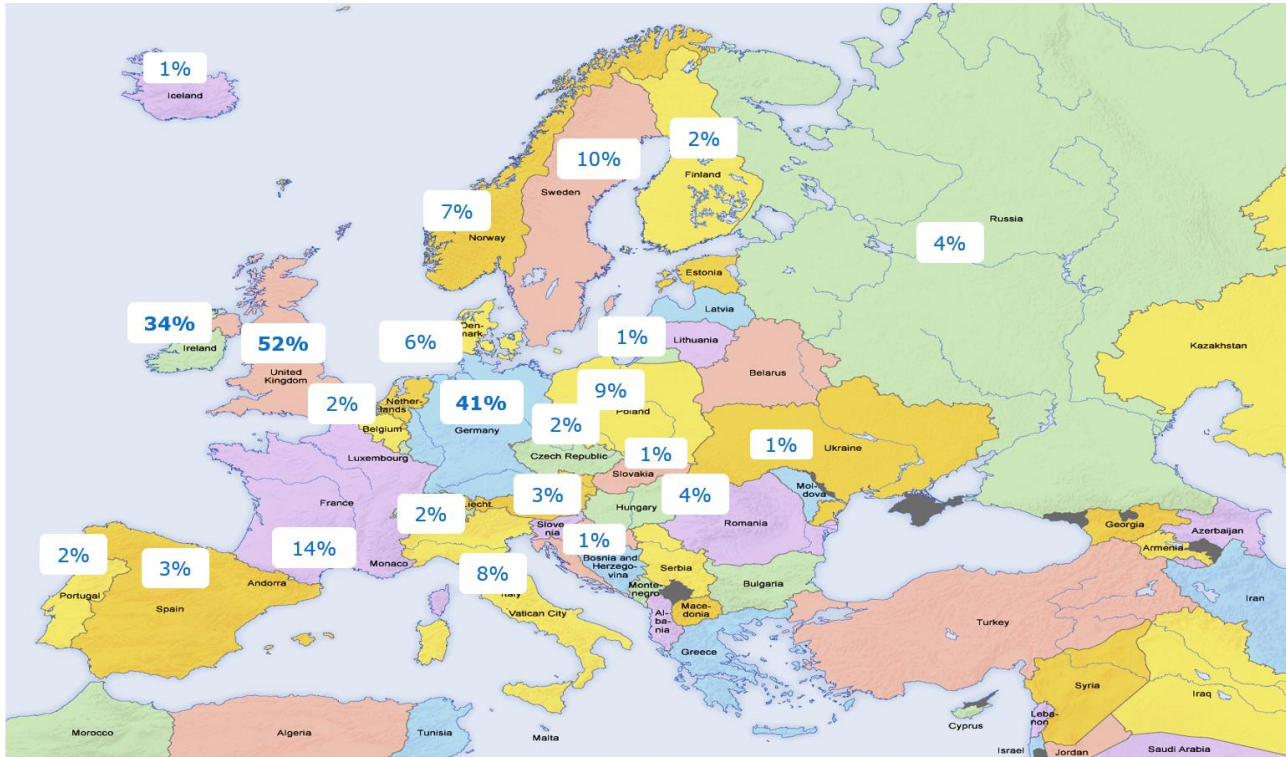
Age when experienced first symptoms with breathing



Q13. What age were you when you first experienced symptoms?
Patients with idiopathic subglottic stenosis n=508

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

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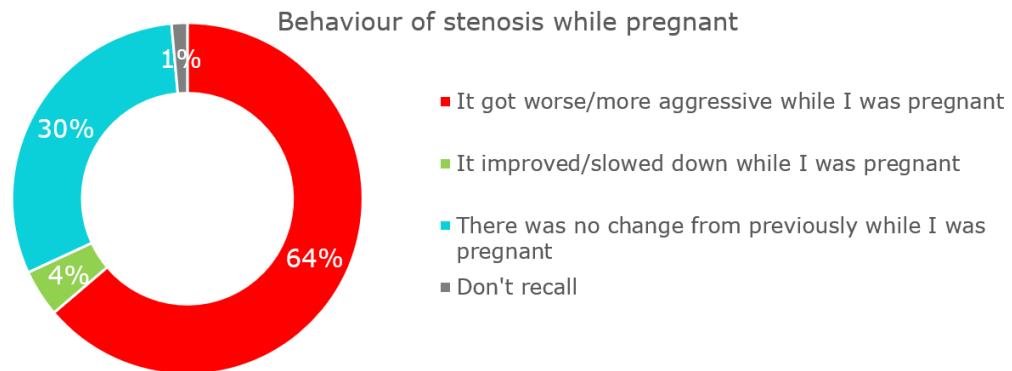
Hormones

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

17% of women with iSGS have never been pregnant, and only one in five (22%) has ever been pregnant while they had iSGS. 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.

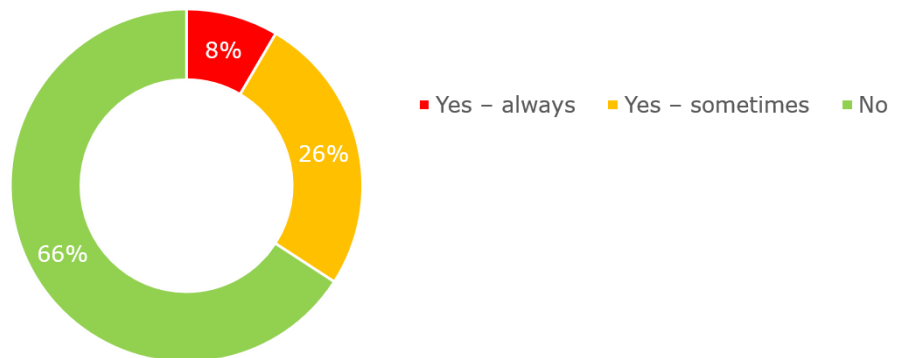
For those women who already have a stenosis when they get pregnant, three in five (64%) see the stenosis get worse during their pregnancy. One in three (30%) see no change.



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*.

Worsening breathing around time of period

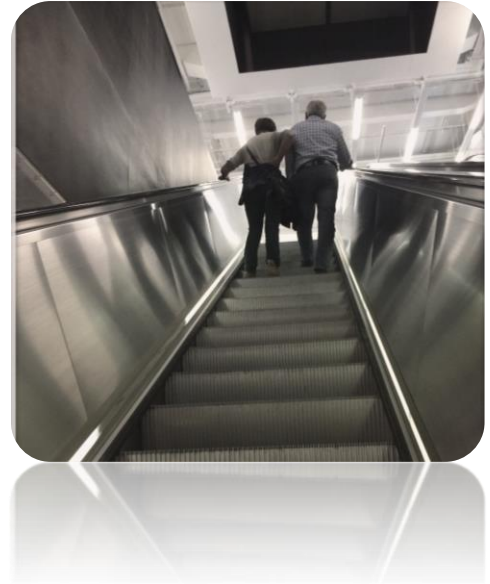


*Journal of Women's Health VOL. 25, NO. 9 | The Association of Inflammation with Premenstrual Symptoms - Ellen B. Gold, Craig Wells, Marianne O'Neill Rasor - Published Online: 1 Sep 2016 <https://doi.org/10.1089/jwh.2015.5529>

Debunking the myths about iSGS

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

"iSGS is caused by reflux" – No. Your iSGS is NOT caused by reflux. Think about it. About 60% of the world's population has reflux – males as much as females – and yet iSGS only (well 98% of the time) affects women. Reflux is not the cause. It **could be an irritant** however. If you think this is the case, talk to your doctor about 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 don't 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 iSGS have more mucus than 'normal people'" – the fact is we generally have exactly the same amount of mucus as everyone else. The difference is that the cilia (little hairs in our trachea) which normally move the mucus up and down our airway with ease have been interrupted by the scarring. This means we have to cough it past the scar. Sometimes mucus can build up and thicken behind the scar and form a mucus plug. This can be quite scary as it can block your narrow airway until you cough it out. Check out our tips to help avoid this, and ensure you **have regular dilations or steroid injections** to avoid the potential of a plug you cannot shift.

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

Many people with this condition run, walk, 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 iSGS.

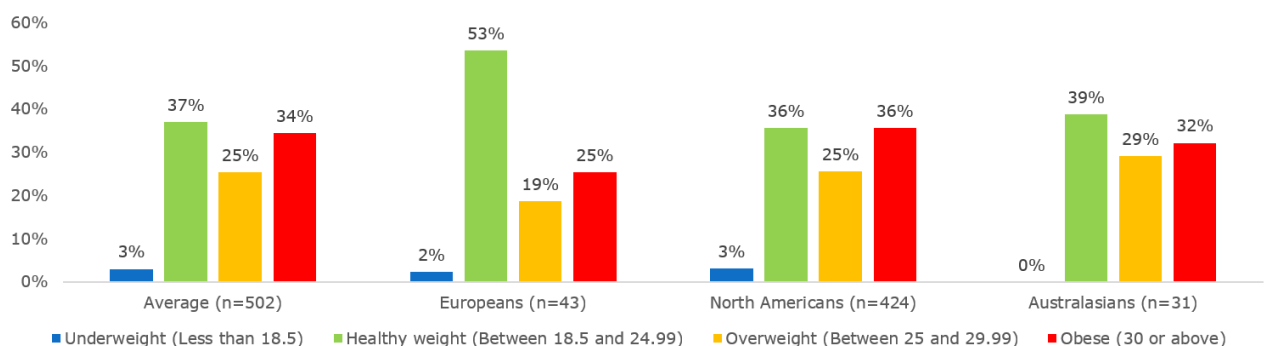
We suggest you start exercising as soon as you're breathing well after a dilation and keep going as long as you can. Just do what you can within your limits and any other conditions you have.

"I am overweight because of iSGS" – again, most of the time this is an excuse. Most of us are overweight because we eat too much of the wrong type of food. Again, this is terrible for your overall health – you are putting stress on all your organs, and your iSGS is not helping. Work hard to reduce weight naturally – cut back on sugar and processed foods and replace with fresh vegetables. Monitor your calorie intake using apps or websites such as My Fitness Pal and become an overall healthier person.

Body Mass Index (BMI) helps us to understand the general health of people by looking at their weight versus their 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 while under anaesthetic– especially if they have had symptoms of sleep apnoea or snoring.

One in three (34%) iSGS 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 research¹ has also found that iSGS 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/>

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

“You must not drink alcohol or caffeine with iSGS” – 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 iSGS 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 (eg resection, Maddern) will cure me of iSGS” – Unfortunately there is no cure to iSGS 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.

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 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
- ANCA (also known as: ANCA Antibodies; c-ANCA; p-ANCA; Serine Protease 3; MPO; PR3; Anticytoplasmic Autoantibodies; 3-ANCA; PR3-ANCA; MPO-ANCA) - test for certain autoimmune disorders, such as Granulomatosis with Polyangiitis (GPA – formerly known as Wegener's granulomatosis (WG)), microscopic polyangiitis (MPA), and a number of others.



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, check the ingredients of Gaviscon reflux medicine, available at most pharmacies)

Summary: what medical interventions are available?

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

Surgical procedures	Medical therapies	Complementary therapies
<p>ENDOSCOPIC (Using a flexible tube, inserted via the nose or mouth)</p> <ul style="list-style-type: none"> Endoscopic tracheal dilation/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, antireflux) Endoscopic tracheal reconstruction (Maddern technique) <p>OPEN ANTERIOR NECK SURGERY</p> <ul style="list-style-type: none"> Cricotracheal resection Laryngotracheoplasty: <ul style="list-style-type: none"> Cartilage tracheoplasty Slide tracheoplasty R-E-A-C-H-E-R (Retrograde, endoscopically-assisted cricoid hypertrophic epithelium resection) Tracheostomy (rare) 	<p>ANTIBIOTICS to tackle bacteria which may be causing inflammation</p> <ul style="list-style-type: none"> Bactrim, a sulpha drug Azithromycin Erythromycin, if allergic to sulpha drugs <p>CORTICOSTEROIDS to inhibit fibroblast formation and reduce inflammation</p> <ul style="list-style-type: none"> Oral tablet (e.g. prednisone) Oral inhalation (e.g. nebulized albuterol, budesonide, eg., pulmicort) Injected (eg, triamcinolone, ie kenalog, between dilations in-office or "awake") <p>PROTON-PUMP INHIBITORS (PPIs) to reduce gastric acid reflux (GERD) if this is an issue for you'. It is recommended PPIs are taken for no 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</p>	<p>HYDRATION to keep mucus thin and slippery (not thick and sticky) and to keep vocal folds moist</p> <ul style="list-style-type: none"> Internal hydration: <ul style="list-style-type: none"> Drink plenty of water Reduce caffeine and alcohol External hydration: <ul style="list-style-type: none"> Steam inhalation Nebulisation with sodium chloride solution Warm air room humidification Nasal rinse Nasal spray <p>MUCOLYTIC THERAPY to help expel mucus</p> <p>Mucolytic medication:</p> <ul style="list-style-type: none"> OTC expectorant cough syrup (eg, guaifenesin) Throat lozenge with glycerin, pectin or slippery elm (not menthol or eucalyptus) Amino acid (N-Acetyl Cysteine, NAC): <ul style="list-style-type: none"> OTC oral capsule Oral inhalation (nebulized NAC, ie, Mucomyst) <p>Airway Clearance Technique or Device:</p> <ul style="list-style-type: none"> 'Huff cough or 'huffing exercise Flutter valve, eg, Acapella <p>LIFESTYLE CHANGES</p> <ul style="list-style-type: none"> Dietary change, anti-inflammatory diet, anti-reflux diet Weight loss Vitamins, herbs Probiotics Enzymes (eg, serrapeptase, bromelain)

Note:

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

Tips to help reduce mucus and coughing

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

Diet

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

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

Food to eat

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

Food to avoid

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

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

Habits to stop

- Smoking! (surely nobody smokes with iSGS)
- Drinking alcohol without enough supplementary water

Drinks

- Plain old water and lots of it
- Tall glass of warm water (non iced) with 1/4 fresh lemon squeezed into it - drink slowly over 30 minutes or so

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

Vitamins & herbal remedies

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

Other solutions

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

Over the counter medicines

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

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.

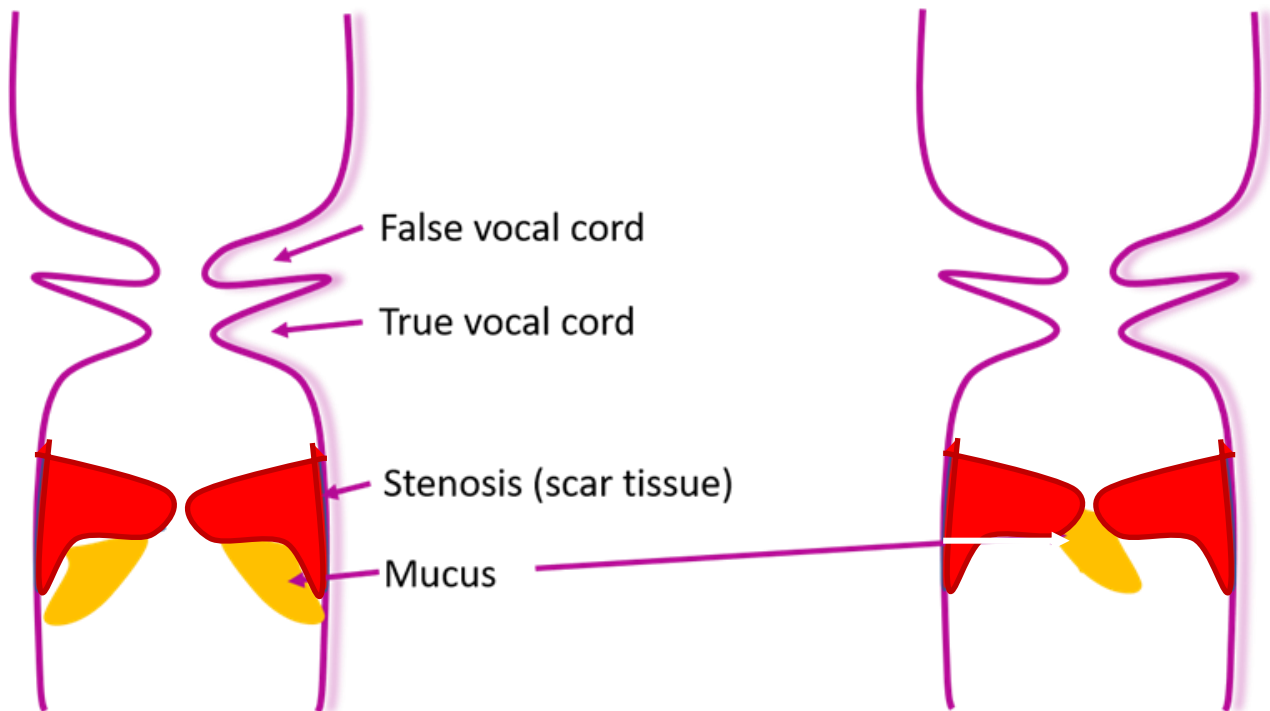
Huff Coughing: Huff coughing, or huffing, is an alternative to deep coughing if you have trouble clearing your mucus. Take a slow deep breath that is slightly deeper than normal. Use your stomach muscles to make a series of three rapid exhalations with the airway open, making a "ha, ha, ha" sound. Follow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

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 doesn't 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 are able to 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 (eg 911/000/999 depending on the country they are in) – it's 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 situation. 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 take action. 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 very 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 iSGS patients have died from this.

Should I be worried about my mucus?

Not all mucus 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. This may or may not be treatable with antibiotics. 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. Can be a risk for plugging, so vigilance needed to keep it moving with fluids.

Tips to help reduce inflammation

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

Food

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

Vitamins and minerals

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

Beverages

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

Over the counter (nonprescription) medicines

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

Your essential medical emergency kit

The next section details what **equipment will help on a daily basis**, 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're travelling and don't have access to your usual doctor(s). It's 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 don't want to have to wait for appointments or calls to be returned.

Vaccinations

Two vaccinations (both via injection) are strongly recommended to help reduce chance of breathing issues:

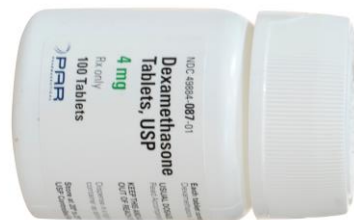
- **Flu vaccine** - the flu can be very serious, leading to complications such as pneumonia, myocarditis (inflammation of heart), neurologic conditions and other bacterial infections – all high risk to airway stenosis patients.
- **Pneumococcal vaccine** - helps protect you against pneumococcal disease. Pneumococcal can cause a fever, pneumonia, meningitis, blood poisoning (septicaemia), ear infections and other airway infections
- **Covid-19** – when available

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's really 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 airway surgeon that you can quickly fill.



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

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



Oral steroids: 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.

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 worsens reflux), and instead the tablet travels further and is absorbed in the small intestine.

If they are not available, ensure you are also prescribed some anti-reflux medicine (such as Somac or similar) to prevent stomach acid reaching your airway and causing more issues.



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.

What equipment will help me?

1. Nebulizer

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

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

How do I use it

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

Why is it useful?

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

We cannot emphasise enough the importance of nebulising when you need to – in the 10+ years the Facebook support group has been running, we have lost three members (that we know of) due to mucus plugs which blocked their airways. There have been multiple near-misses with other members. Nebulising can help you to avoid a life-threatening situation.

What types of nebulizer are there?

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

Omron MicroAir – portable nebulizer

Approx. \$170-\$200 online



- Small and light – handbag sized
- Silent
- Portable – can run without mains power so can use in the car, watching tv, doing the housework or cooking
- Runs on batteries (last around 4 hours)
- Some delicate parts so care needs to be taken when using and cleaning

Allersearch Portable Nebuliser

Approx. \$160-\$180 online



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

Other brands Eg Allersearch Ventalair Max

Approx. \$210 online



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

2. Humidifier

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

How do I use it

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

Why is it useful?

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

What types of humidifier are there?

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

Fisher & Paykel My AirVo 2 system

\$3,000 - \$4,000 – may be available to rent



- Individual high-flow warm humid air into the upper airway
- Aids in liquefying debris in the upper airway and help patients cough that material out or swallow
- Similar to what you would get in hospital to help you clear mucus in your airways
- Expensive to buy

Vicks Warm Steam Vaporizer

Approx. \$50 online new



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

3. Medic alert bracelet

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

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

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>

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

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

Suggested wording: Subglottic stenosis; very narrow airway, use paediatric tube

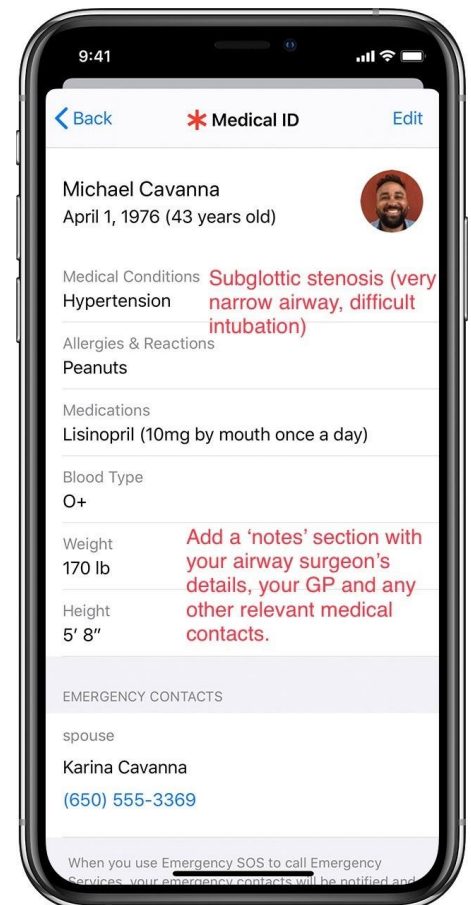
4. iPhone users: *Medical ID

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

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.
- 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.



5. Peak Flow Meter

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

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

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

How do I use my meter?

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

How often should I use my meter?

Ideally once a day.

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

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

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

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

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

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

My numbers are declining. What does this mean?

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



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

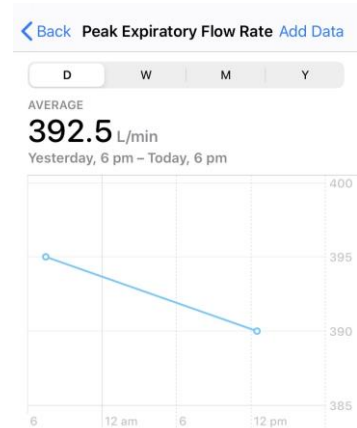
Peak Expiratory Flow (PEF) or peak flow readings vary widely between individuals and devices.

Where should I record my values?

If you are part of the NOAAC study, you can record PEF results using the Airflo app (<https://noaac.net/tools/>) or manually on a peak flow chart. Another app you might use is the AsthmaMD app – which will also allow you to chart your readings: <https://www.asthamamd.org/>. iPhone users will also find a tracking feature under the Health app which comes with the phone.

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

If you have an iPhone, the free 'Health' app that comes with the device has a section called 'Respiratory' which includes a chart for 'Peak Expiratory Flow Rate' recording.



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

The last thing many of us would want to do after surgery is to blow hard on anything, so, no, give yourself a few days to recover. Resume use of your peak flow meter when it feels comfortable to do so. 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. All evidence is essential to help your specialist offer advice about any potential next steps in your care.



6. Flutter valve device

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

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

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

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

*Acapella flutter valves pictured.



What is a dilation or dilatation surgery?

Once you have been diagnosed with iSGS your surgeon will most likely book you in for a dilation operation (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 up 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 up 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 and prevent the scar travelling up towards the vocal cords. Anecdotal findings from doctors in London shows 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 up 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).

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.
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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 definitely 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 commonly 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 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.

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

Your neck and back may feel stiff and sore from the positioning of you during surgery. The longer the surgery, the more prone you will be to 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).

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

What medication am I likely to be on afterwards?

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

Some patients are also prescribed an antibiotic (such as Bactrim) - they may be on this long term. The antibiotic is thought to help by controlling bacterial infections at the site of the scar. As doctors dilate, the tissue becomes damaged and dies off. It is though that bacteria may then colonize the wound and create further damage. It is a relatively benign medication which would be OK to be on for a while (months to a couple of years) but it is not recommended to take it long term for this disease.

How long before I should go back to work?

Usually recommend about 3 days away from work to rest post operation, with at least 2 days resting the voice post op. Some doctors recommend up to 5 days of no talking. 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 wouldn't advise running or anything too strenuous. Basically, go with gut feel. The anaesthetic will make you feel sluggish for a day or two, after that you should be able to go back. Best advice: listen to your body - if you don't feel like it, then don't do it! Don't over stress yourself and put back your recovery.

Will I only need one dilation?

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

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

A tracheal resection is a complex surgery that involves removing a scarred portion of the trachea and reconnecting the healthy ends. You may hear it also called Cricotracheal resection (CTR), Laryngeal Tracheal Resection (LTR) or Tracheal resection and reanastomosis. 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. Recent research among iSGS patients finds that 73% of patients believe their resection was reasonably or very successful. Around one in five (18%) found their breathing more of a challenge after the operation – needing dilations again or another major surgery. 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?

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

What exactly are the risks?

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 restenose 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 is 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 mentions they are planning 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 reasons for using this and their expertise.**

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 confident with the level of experience of your surgeon, then you should be safe with this technique.

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

If your doctor wants an alternative, point them towards the website created by experienced airway surgeon Dr. Paul Castellanos – www.airwaysurgeon.com – there he shows an alternative method.

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

How bad is the scar?

The scar is about 3 inches in length.

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

What is The Maddern Technique (endoscopic tracheal reconstruction)?



How is The Maddern Technique different to a resection?

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

This operation is endoscopic rather than via open surgery meaning the neck is not cut open. This has several benefits including a minimal risk to the vocal 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 don't bleed too much and your airway can cope well with a size 12+ open stent then you are unlikely to have a tracheostomy.

When can I get up and walk around?

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

When can I eat and talk normally?

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

How long am I in hospital?

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

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

Will I be in pain?

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

How long until I feel better post op?

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

Some people may still have some difficulty with mucus – after the surgery you will need two 'tidy-ups' – these are to ensure the all graft is removed and 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's important while in hospital to ensure the original dressing is left on and kept dry to aid the healing process. Tape a rectangle of plastic bag over the site when showering to protect it.

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

The first patient to have this operation in 2012 is still breathing well. It is a new operation so there are no long-term case studies or statistics, but most patients so far are doing well.

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

What is the REACHER?

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

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

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

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

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

What are in office/awake steroid injections

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

Why would I choose to have a steroid injection?

Inflammation is a key part of iSGS and often causes major issues with our breathing, even when the actual scarring is not too bad. Taking regular oral steroids can help, but have many negative side effects. A 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. 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. Given a choice, many suggest internal injections are less painful than external injections.

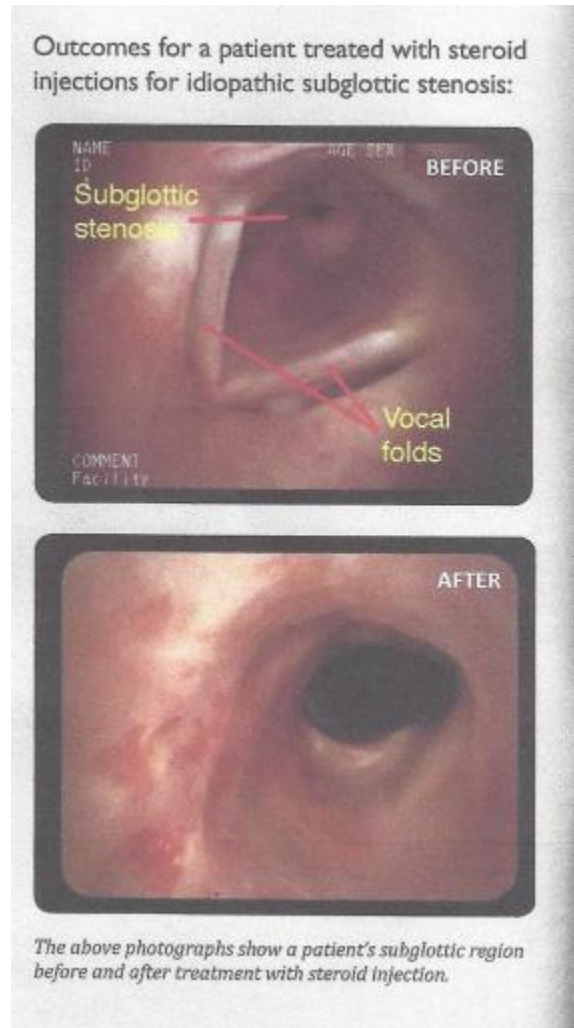
Will it affect my voice?

Some have a short-term huskiness while the vocal 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.

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



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:

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

What are the risks with this protocol?

As with all drugs, these can have side effects.

- Antibiotics: Some people can have negative side effects with taking antibiotics long term, such as thrush and stomach issues. Some people cannot tolerate sulpha based drugs, in which case an alternative is found.
- Steroid inhaler: This can cause oral thrush – you need to wash your mouth out and gargle with water after you take your breath. Some people can find the powder will coat their vocal 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

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.

What questions should I ask my surgeon?

At **every appointment** with your specialist:

What is the current diameter of my stenosis in millimetres?	<i>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).</i>
At which percentage is my airway closed?	<i>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).</i>
What is the current distance of the top of my stenosis from my vocal cords?	<i>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.</i>
How has my stenosis changed since my last appointment?	<p><i>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?</i></p> <p><i>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.</i></p>
What are my ANCA test results? And when were they last tested?	<i>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.</i>

Do both my vocal cords work as they should?	<i>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.</i>

When having **dilation surgery**:

How long should I expect to be in hospital?	<i>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.</i>
What can I do to protect my teeth from damage?	<i>Many hospitals will insert a mouth-guard 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.</i>
What surgery are you planning to perform? A laser dilation? Balloon? Coblation?	<i>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 doesn't do this operation.</i>
Will you be doing any steroid injections? Applying Mitomycin-C?	<i>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.</i>
Are you planning to do a biopsy of the scar to test for ANCA? Or take bloods to test for this?	<i>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.</i>

When considering **major surgery**:

How many resections/reconstructions have you done? How many on idiopathic patients? How many on non-idiopathic?	<i>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.</i>
What is your success rate? How do you define success?	<i>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?</i>
What do you see as the general risks of this surgery?	<i>For example Restenosis? Nerve damage? Infection? Trach? Weak voice? Unable to sing?</i>
What do you see as my personal risks?	<i>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.</i>
How can I personally reduce my risks of complications?	<i>For example this may mean gaining or losing weight, cutting out bad habits, doing pre-operative exercises or stretches.</i>
How long will I stay in the hospital?	<i>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.</i>
Can I talk to any of your former patients?	<i>Most experienced surgeons will have patients who are willing to share their surgery story and help you decide whether or not you are comfortable proceeding with the surgeon and procedure.</i>

What is your major complication rate? What major complications have you experienced?	<i>This will both help you prepare for the worst-case scenario, and also 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.</i>
What happens during the operation? How long does it last?	<i>The more you understand in advance, the more comfortable you will feel about the surgery.</i>
What can I expect when I wake up? (eg lines, drains, masks, catheter, feeding tube)	<i>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.</i>
How will I feel when I wake up? (Pain? If so where)	<i>Understand more about managing expectations and how the recovery team will be managing this – through pain killers or muscle relaxers, steroids and so on</i>
When will I be able to eat and drink normally?	<i>For example moving on from ice chips to solid food or having a feeding tube removed.</i>
How long before I am allowed to go home?	<i>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.</i>
What sorts of complications might I encounter after leaving the hospital and how should I deal with those if they occur?	<i>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?</i>
Will I have any restrictions once I go home?	<i>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.</i>

Am I likely to need to take medication once I am discharged? For how long?	<i>These could include steroids, anti-reflux medication or antibiotics for example.</i>
Is there any equipment I would need to use at home?	<i>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 (eg in heating or air conditioning) easier to breathe.</i>

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!).

Going in for a day

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



Going in for a night

Same as going in for a day plus:

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

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

Same as going in for a night plus

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

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

Tips on coping 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 the 5:2 diet for a year before their op - this reduces your sugar levels, bad cholesterol and puts your body into repair mode as well as weight loss. Ensure your body is working at its most efficient to help repair the surgery site.

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

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 - eg - being allowed to use the bathroom rather than a bedpan, getting your feeding tube removed, being allowed to wash yourself, then shower...even taking fewer pain killers is progress...

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

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

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

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

Sleep - Sleep is a major challenge in hospital. Not only are there nebulisers and machines making

noises all over the place, alarms and nurses chatting, but you're also woken for pain meds and blood pressure and so on.

Having some noise cancelling earphones is essential – eg Bose ear buds block out 80% of all sound even without playing music, just with the power on. If someone wants to buy you a 'good luck in your operation' present this is what you want!

Eye masks are the other solution. 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 you 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're being given and why. Again this is about empowerment and also interesting. Means that you're part of your treatment rather than just experiencing it.

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

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

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

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

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

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

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 6.0mm *[smaller if your airway doctor recommends]*
- ETT cuff pressure measurements (suggested 20-30 cm H2O 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 iSGS - many patients have successfully had children with this disease.

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

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

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

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

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

Airway Surgery During Pregnancy

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



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

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

Frequently Asked Questions

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

Airway stenosis in pregnant iSGS patients can be treated in the office with awake, 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 (ie. intravenous or oral) and inhaled steroids (both nasal and pulmonary) have an established track record in pregnancy (particularly in asthmatic patients), and are widely regarded as safe in pregnancy.

Do I really need surgery during pregnancy?

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

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

Will the anaesthetic and drugs harm my baby?

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

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

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

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

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

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

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

Will I need to be intubated to deliver my baby?

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

Tips for dealing with iSGS during pregnancy

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

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

Remember to include details of your doctors and how to contact them.

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

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

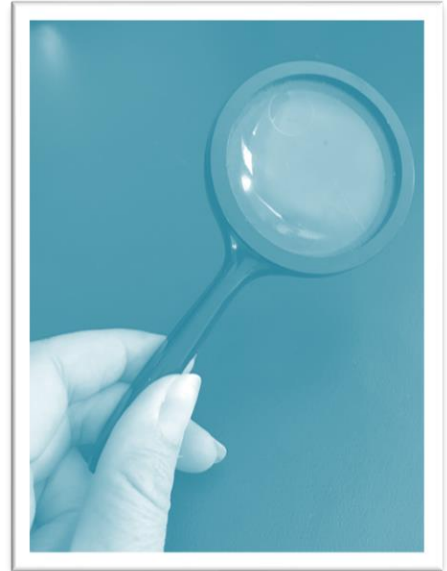
Try to relax! While it can be stressful and scary dealing with this condition during pregnancy - try not to get worried about it.

Research in progress

Research you can participate in now:

- **Global study:** A major study to *understand the treatments and efficacy of treatments for iSGS* is being carried out by Vanderbilt University in Nashville Tennessee
- **Global study:** Research into patients with idiopathic subglottic stenosis who have *a blood relative also with the disease* (e.g. mother, sister, grandmother, aunt, cousin)
- **USA & Canada only:** A study looking at patients with iSGS who had their first symptoms under the age of 35 years

Email Cheryl if you are interested in taking part in any of these three studies: cheryl.kinnard@vumc.org



To participate in future studies:

If you are willing to take part in other upcoming research regarding iSGS, share your email address with Catherine via idiopathic.subglottic.stenosis@gmail.com

Other research

- **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 at the University of Miami are setting up a study soon looking at the use of hormones and iSGS
- **Global:** Doctors at Ohio University are partnering with Living with Idiopathic Subglottic Stenosis group to survey patients about their experiences with in-office procedures and the recent Covid-19 pandemic
- **Global/USA:** A PhD student is preparing a thesis about iSGS patient experience with the aim of helping to decrease the time to diagnosis. Research in particular is coming up for those iSGS members who are also GPs/primary care physicians
- **Global/UK:** Research to improve clinic processes with iSGS patients to ensure they are more streamlined, patient led, relevant and address all patients' concerns

Latest findings

Inflammation and bacteria

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

Successful surgeries

A good proportion of this research has been about proving what we already assume – that dilations (balloon or laser or a combination) are a short term solution. The study has definitely proven that, observing that on average just over 1 in 4 patients will need another dilation within 3 years (28%). A dilation, however, is seen to have very few downsides, with a small percentage of patients experiencing some short term tongue numbness and a small number experiencing dental damage.

This is then compared to an open resection surgery. The positive of this is that within a 3 year period the patient's stenosis is unlikely to return (only 1% chance within 3 years), but the downsides are quite a few, including damage to one or both vocal cords (and hence permanent damage to voice) and other post op issues with neck pain and so on.

A newer style of endoscopic resection surgery was compared. Mayo in Rochester has the most experience with this, but it also is undertaken in Maryland (Dr Hillel) and Tennessee (Dr Gelbard), and may also be conducted elsewhere. At a basic level, it involves removal of the scar leaving gaps in the mucosa, then post op the patient uses a Flovent/Pulmicort inhaler plus bactrim antibiotics and reflux medicine if there is evidence of that – I'm sure it's way more complicated than that, but much less so than an open tracheal resection. This surgery has very few downsides, and within three years, only 12% of patients had their stenosis reoccur. This operation is endoscopic- meaning no big scar or the risks to important nerves and voice. It could end up as the first port of call instead of a dilation, with much longer lasting results. (For more information on this technique see: Idiopathic Subglottic Stenosis: An Evolving Therapeutic Algorithm

Laryngoscope, 124:498–503, 2014 – you can find this paper under the 'Files' tab in the group). More testing is needed to compare success rates with and without the Bactrim.

The final published report can be found here:

<https://jamanetwork.com/journals/jamaotolaryngology/fullarticle/2753766>

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 would be 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 Maldonado

Dr Jan Kaserbauer, Dr Eric Edell and Dr Dale Ekblom

Dr Robert Lorenz, Dr Paul Bryson and Dr Michael Benninger

Dr Marshall Smith

Dr James Damiero

Dr Donald Donovan

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Mr Guri Sandhu

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Dr Joshua Schindler

Dr Paul Castellanos

Dr Phillip Weissbrod & Dr Samir Makani

Dr Mark Courey

Dr Matthew Clary & Dr Daniel Fink

Dr Gregory Postma

Dr Henry Hoffman

Dr Andrew J McWhorter

Dr Ramon Franco

Dr Robbi Kupfer, Dr Norman Hogikyan and Dr Robert Morrison

Dr Stephanie N Misono

Dr Christopher M Bingcan

Vanderbilt University

Mayo Clinic (Rochester)

Cleveland Clinic

University of Utah

University of Virginia

Baylor College of Medicine

John Hopkins

Imperial College London, UK

Mayo Clinic (Scottsdale)

Oregon Health Sciences University

University of California, San Diego

University of California, San Francisco

University of Colorado

Augusta University

University of Iowa

Louisiana State University

Massachusetts Eye and Ear Infirmary

University of Michigan

University of Minnesota

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	Landspítali 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
Mr Reza Nouredine	Poole Hospital NHS Foundation Trust & University of Southampton

Glossary of terms associated with subglottic stenosis

iSGS 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), is a patient airflow monitoring tool (<https://noaac.net/tools/>) that measures and records 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 very 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 iSGS 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 iSGS 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 laryngotracheoplasty.

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. iSGS 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 (eg Prednisone), by oral inhalation (eg nebulized albuterol, budesonide, ie Pulmicort) and by an injection (see Steroid injection and Awake injections). Common corticosteroids used in iSGS 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 anesthesia, 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 anesthesia.

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 dyspnea, 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 innerpart 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, ie 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. iSGS patients may wish to add their recommended ETT size to a Medical Alert ID.

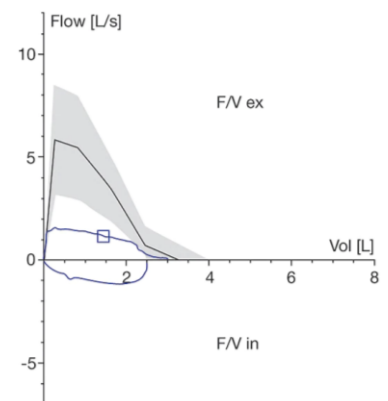
Erythrocyte sedimentation rate (ESR): ESR is a blood test for inflammation.

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. iSGS 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 autoimmune collagen vascular disease.

I

Idiopathic subglottic stenosis (iSGS 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. iSGS 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 iSGS patients helps us understand the molecular pathogenesis of iSGS 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 anesthesia, 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. iSGS 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 iSGS 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 glycerin, pectin or slippery elm not menthol or eucalyptus; amino acid N-Acetyl Cysteine (NAC) or NAC as a capsule or nebulized NAC, eg 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 iSGS 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 iSGS 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, ENTdoctors, 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 (ie the force of airflow through the bronchi and thus the degree of any airway obstruction). It gives you a numeric reading on a scale; peak flow readings are higher when the airway is open, and lower when the airway is narrowed (constricted).

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 iSGS 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.

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 anesthesia 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 nonprofit 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 iSGS 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)

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