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Idiopathic Subglottic Stenosis
A Rough Guide for Beginners

Last updated: 12 April 2016
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 and 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 our Facebook Group. Hopefully it didn’t take too long. We understand where you are coming from and hopefully this document will help.

So back to the question in hand. What is idiopathic subglottic stenosis (iSGS from hereon. All mentions of iSGS also apply to other forms of stenosis)?

Getting down to basics;

- **Idiopathic** – is a fancy way of saying ‘no known cause’
- **Subglottic** – is below the glottis area of your trachea. You may have been told you have tracheal stenosis – this is a more general description detailing that your stenosis is located within the trachea
- **Stenosis** – another fancy word for ‘narrowing’

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

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

**Idiopathic** – Doctors suggest your stenosis is idiopathic if you have no history of significant laryngotracheal injury. No significant history of endotracheal intubation or tracheotomy within 2 years of your first symptoms. No thyroid or major anterior neck surgery. No exposure to radiation on your neck. No caustic (eg chemical burn) or thermal (heat) injuries to the laryngotracheal area. No history of vasculitis (a condition that involves inflammation of the blood vessels). Negative test results for angiotensin - converting enzyme (ACE) and antinuclear cytoplasmic antibody (ANCA) – your doctor should have done tests for these. The lesion must involve the subglottis.

**Autoimmune** - Patients with documented clinical along with serologic and/or histologic diagnosis of Wegeners (GPA), Relapsing Polychondritis (RPC), Systemic Lupus Erythematosus (SLE), Rheumatoid Arthritis (RA), Epidermolysis Bullosa (EB), Sarcoidosis, or Amyloidosis. Again, your doctor should check for these.

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

**Iatrogenic** - Patients that developed subglottic or tracheal stenosis following tracheostomy. Or subglottic or tracheal stenosis developing within 2 years of intubation.
What are the common symptoms of iSGS?

We all share some common symptoms:

**Stridor** – this is what we often will describe as a wheeze, the sound when we breathe. It will often be silent most of the time, but get worse under exertion or in stressful situations. When your stenosis is particularly narrow, you may hear your stridor all the time. 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 due to the little hairs on your trachea being absent where the scarring is. These hairs usually help move mucus smoothly up and down your airway. Where they are missing, you will need to cough the mucus past. The coughing will often get worse when you’re talking lots, exercising or in stressful situations.

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

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

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

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

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%. It is estimated that this condition impacts one in a half a million people. If that is true, this group only contains a fraction of those out there.

Based on that statistic there would be an estimated:

<table>
<thead>
<tr>
<th>Country</th>
<th>Patients</th>
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<tbody>
<tr>
<td>USA</td>
<td>630</td>
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<tr>
<td>Brazil</td>
<td>400</td>
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<td>Germany</td>
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<td>New Zealand</td>
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<td>South Africa</td>
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Perhaps many have not yet been diagnosed, while some will have been resected or reconstructed and have no need of a support group. Others will not be on the internet or speak or write English.

If you have a relative with this condition then you are extremely rare – only 1% has a relative with this – usually a sister or mother.

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

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

One question asked frequently is where do we all live. Research conducted among group members in 2013 showed that the majority live in North America (including the USA, Canada and Mexico), followed by Europe (including the UK, Germany, Sweden, Iceland and The Netherlands), Australasia (Australia and New Zealand) and a rare couple from Asia (India and the Philippines), South America (Brazil) and Africa.
This is almost certainly skewed by the fact this is an online group and was an online survey (therefore requiring computer access) as well as the fact it is in English.
Debunking the myths about iSGS

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

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

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

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

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

There are plenty of alternatives out there if you look – these days you can get electric bikes which give you some exercise while helping you on the hills, try kayaking with 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 as soon as you’re breathing well after a dilation and keep going as long as you can. Just do what you can within your limits and any other conditions you have.

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

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

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

- ESR (test for acute inflammation)
- CRP (test for acute inflammation)
- Rheumatoid factor (test for autoimmune disease)
- ANA (test for autoimmune disease)
- C-ANCA (test for Wegener’s granulomatosis)

Your doctor may also take a biopsy while under anesthetic which will be tested.
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
- 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.

Food to avoid

- Dairy products
- Wheat & Gluten
- Soy
- Sugar

Habits to stop

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

Drinks

- Plain old water and lots of it
- Tall glass of warm water (non iced) with 1/4 fresh lemon squeezed into it - drink slowly over 30 minutes or so
- Herbal teas with a slice of lemon in - drink slowly
- Coltsfoot tea (though read up about it first and only drink for max of 4 weeks at a time)
- Soda water (it breaks up mucus so it’s easier to cough up)
- Thyme (you can make tea with it) clears secretions. In Europe there are thyme teas and extracts for bronchial symptoms
- Pineapple juice mix - 1 Cup fresh pineapple juice, 1/4 Cup fresh lemon juice, 1 piece fresh grated ginger (about 1 & 1/2 TBS), 1 TBS Raw Honey, 1/2 tsp cayenne pepper. Mix all
ingredients in a blender or juicer or stir in a glass until well blended. Drink about a 1/4 cup 2 to 3 times per day.

- Lemon verbena is also effective at loosening up and removing mucus from the lungs and sinus passages

**Vitamins & herbal remedies**

- Echinacea
- Goldenseal (also known as Golden Seal)
- Slow release vitamin C/Ester C 2000g
- Thyme

**Other non-chemical solutions**

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

**Over the counter medicines (manmade solutions)**

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

Many of us have inflammation in our airway - this can be caused by excess coughing but can be eased. There are a number of 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
- Apples and cherries are also anti-inflammatory
- Fresh ginger - try it in tea or stir fries
- Lemon - half a slice in tea, half a slice in water
- Eat fresh food you make yourself - avoid processed foods and high sugar
- Don't use vegetable oil in cooking - switch to extra virgin olive oil, grape seed, coconut or avocado oil
- Eat more fish rich in omega 3 - minimise red meat
- Snack on nuts and seeds - almonds, hazelnuts, walnuts, sunflower seeds in particular
- Fresh garlic - best uncooked!
- Herbs - especially mint (try chopped up in salad), basil, chili peppers, oregano, parsley, rosemary, thyme, and the wonderfully powerful turmeric.
- Dark chocolate (70%+)

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

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

Over the counter (nonprescription) medicines
- Difflam (may have other names in different countries - this is the Australian brand name) Anti-Inflammatory, Anti-Bacterial Lozenges - helps to reduce inflammation and coughing and mucus
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 fairly noisy and involve putting a mask over your face and breathing in. In 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!
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**
- 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 off batteries (last around 7 hours) or mains power (purchased separately – plug available for all countries)
- Some delicate parts so care needs to be taken when using and cleaning

Approx. $170-$200 online
Power adaptor add approx. $25

**Allersearch Portable Nebuliser**
- 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

Approx. $160-$180 online

**Other brands**
**Eg Allersearch Ventalair Max**
- 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

Approx. $210 online
2. Humidifier
A humidifier is a device you fill with water, turn on and it puts moisture back into your atmosphere. Particularly useful where you have air conditioning or heating on where the air is particularly dry.

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

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

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

- Vicks Warm Steam Vaporizer
  - It does what it says on the box
  - Suitable for a mid-sized room
  - It is hard to clean
  - Some report it trips power

Approx. $50 online new

3. Pulse Oximeter
A device to check your oxygen saturation

Why is it useful?
It measures oxygen levels in your blood and allows you to monitor what is ‘normal’ versus ‘abnormal’ for you. Your oxygen levels should be 90% or above.

What types of oximeters are there?
There are many on the market. You should easily be able to find them online for around $35
4. A medic alert bracelet

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

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

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

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

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

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

Suggested wording: Tracheal stenosis; difficult airway intubation; use fiber optics
5. iPhone users: *Medical ID

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

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

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

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

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

After the Medical ID has been created, users can always go back and make changes at any time through the Health app.
What is a dilation or dilatation surgery?

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

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

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

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

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

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

How long does the dilation operation take?

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

How will I breathe while I am being dilated?

The anesthesiologist will intubate you with an airway tube while your doctor takes a look with the scope and gets set up. Your lungs are then hyper-inflated and the airway is pulled out while you are basically holding your breath and the balloon is inserted. While the balloon is in, they are carefully monitoring your blood oxygen levels to make sure they don’t drop. You may have chest pain for a few days after the surgery- the hyperinflation of your lungs may cause this.
What is mitomycin C?
Looking it up online can be quite daunting when you find it is a drug used in cancer treatment (chemo). It does not mean you have cancer or are having chemotherapy. It is applied to your scarring at the end of the dilation in the hope that it will stop the scar cells from growing back. There is no evidence that this works, but most patients do not experience any adverse side effects (you might be able to taste it as a bitter flavour when you cough once you wake up).

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

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

Some people may have a husky voice for a few days post operation - at most for around 10 days - but this will go as any bruising subsides. In a few cases people lose their voice for 1-3 weeks.

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

Some doctors recommend using a humidifier 24 hours per day for a month post-surgery to keep the airway moist so that there is no crusting of the surgical area. A saline nebuliser for 30 minutes a day for a few days post op can also help soothe it.

What medication am I likely to be on afterwards?
Some doctors prescribe Tylenol 2 with Codeine (USA) or Panadene Forte (Paracetamol with Codeine – UK and Australia). This provides pain relief and suppresses any cough for the first few days.

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

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

**How long before I should go back to work?**

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

**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?**

It varies widely. There are some who only have one and no more. Unfortunately these are in the minority. The majority of patients will have a second one within two years. Some can cope more than two years between dilations.
What is a resection (resection and reconstruction) and how is it done?

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

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

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

Can previous treatments 'mess up' your candidacy for resection?
Opinion on this is mixed. Some doctors believe the skin toughens up making it more challenging to work with, while others believe there is no problem. The majority of skilled doctors will still be able to conduct a resection no matter how many dilations you have had. There may be other reasons you are rejected for a resection, however.

What exactly are the risks and the percentages of patients that experience any of these risks?
Main risk is restenosis. Next is damage to the nerves of the vocal cords. About 1% chance of bleeding or infection. Dr. Mathisen said that with his group's 250 resections on iSGS patients- 5% will go on to restenose. 50% will have some change in voice but 90% will be fully functioning- the remaining 10% may have a weaker voice and inability to project. 2-3% will have permanent hoarseness if one of the nerves is damaged. Swelling after surgery is the biggest concern but only 1-2% will need a post-surgical trach temporarily.

How bad is the pain? How long are you on pain meds?
Talk to your doctor ahead of time about pain relief and muscle relaxers. Some say the position the chin stitch puts you in is the greatest pain. The hospital will manage your pain in the best way for you. Some report experiencing a pain level of maybe 7 out of 10 day one and two, 5 out of 10 day three and then maybe a 2 out of 10 for the final few days.
What is a chin stitch?
The chin stitch is to keep your head in a downward position after surgery so that the trachea can heal. It is simple a stitch of thread that goes through the skin under your chin (in one hole and out another) to your chest (near the top of your sternum). More than anything- it is a reminder not to move your head too far so that the trachea can heal up. There is a YouTube that someone recently posted of a resection. At the end of that video you can see them inserting the chin stitch. Also if you look at the photos tab there are some photos that show the stitch running between the chin and chest.

Did you have difficulty swallowing after surgery? How long before you can start to eat and drink?
Most are on a feeding tube for up to 5 days before being placed on a soft diet. You will likely be given medicine via IV to calm your stomach.

How long after surgery before they have you up and around?
You are encouraged to move around and are likely to be walking within the first two days.

I saw mention of a drain, what is that, where is it placed and for how long?
It comes out from your chest or beside your neck incision and it is in 3 or 4 days, it is more unattractive than painful.

Do you need to have an NG tube and catheter? If so, for how long?
You are likely to be given catheter. As long as you can walk they will let you take it out.

Are you on antibiotics or any other type of meds following surgery? If so, how long?
Some receive steroids to keep swelling down as a daily injection at the hospital, and then taken orally on a tapering dose for 2 weeks. You may also be given acid reflux medication as a precaution.

Were you told not to speak for a period of time after surgery? If so, how long?
There are varying experiences – some are asked to speak right away, while others are put on voice rest for several weeks.
How long did it take for your voice to get as good as it was going to get?
This is not known for sure – some have to have voice coaching to strengthen their vocals and return to previous levels. Others will always have a weaker or raspy voice.

What is the recovery time and what can I do during this period?
Minimum of 6 weeks off, and return to work depends on each individual, as everyone feels and recuperates differently. No driving for 8 weeks

How bad is the scar?
The scar is about 3 inches in length.
Can also have a small 1 cm scar where the drain is located
What is The Maddern Technique (endoscopic tracheal reconstruction)?

At the time of writing, this technique is only practiced in the UK by Guri Sandhu at Charing Cross Hospital in London and in the USA by Dr Robert Lorenz in Cleveland, Ohio. As of writing this, more than 20 patients had undertaken this surgery with success to date.

Other doctors in the USA, Canada and Australia have expressed an interest in knowing more about this procedure, which is lower risk than a resection and has a faster recovery time. It is thought by many that this will replace resection surgery in time.

If you think your doctor should be considering this for you, put them in touch with Mr Guri Sandhu: +44 (0)207 486 0022 or by emailing his secretary, Radka Louis: radka@harleyent.com or for those in the USA contact Dr Robert Lorenz, Associate Professor of Surgery, Head and Neck Surgery 9500 Euclid Avenue, Cleveland, OH 44195. Phone (216) 444-8500 to make an appointment.

How is The Maddern Technique different to a resection?

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

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

Broadly what happens in the operation?

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

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

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

The tracheostomy is provided to allow you to breathe while your airway is swollen and the stent is in. You will have this for around 5-7 days. If you don’t bleed too much and your airway can cope well with a size 12+ open stent then you are unlikely to have a tracheostomy.
When can I get up and walk around?
Usually the following morning after your operation you will at least be able to visit the bathroom (you are not given a catheter and the bed pan is not a fun experience!), and you will feel more and more like exploring as the days pass. Once you are off the feeding tube and IV you will be able to make more extensive journeys to the coffee shop and even outside (as long as you let the nurses know when to expect you back!).

When can I eat and talk normally?
Once a speech therapist has visited you and ensured you are able to swallow properly you should be able to have the feeding tube removed. To a large extent 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 a week. 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 lazer ‘tidy up’ the new skin and so your doctor can check out how it is healing. You may need another final lazer ‘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?
Immediately! As soon as you are recovered from the general anesthetic 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.

The graft site on your leg 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 as long as possible to aid the healing process. Tape a rectangle of plastic bag over the site when showering to protect it.

How long does it last? How will I feel a year later?
The first patient to have this operation is breathing well 4 years later. It is a new operation so there are no long term case studies or statistics, but all patients so far are doing really well.
What is The REACHER (Retrograde Endoscopically Assisted Cricoid Hypertrophic Epithelial Resection)?

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

It is similar in many ways to the Maddern 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).

The “R” of REACHER stands for “retrograde”, in which the trachea is opened and the subglottic area is actually approached from below, while an intra-operative breathing tube is used to ventilate the lungs but removed prior to the end of the operation. This allows patients who are not able to be successfully oxygenated with the “JET” ventilator for the length of the Maddern Procedure to still be candidates for these minimally-invasive procedures. Although the operation is technically “open” and not done endoscopically, since endoscopes are still utilized to see better and magnify the operative site, it is called “Endoscopically-Assisted”.

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

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

There is some expectation that the Maddern procedure will become the standard with the REACHER reserved for patients with either hard to visualise subglottic areas through the laryngoscope, or else patients who cannot tolerate jet-ventilation (their oxygenation cannot be maintained throughout the endoscopic procedure).
I am considering major surgery. What should I ask my surgeon?

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

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

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

**Going in for a night**
Same as going in for a day plus:
- Wash (toiletries) bag - definitely want to clean your teeth, possibly your own soap
- Tissues
- Nighttime routine things - face creams and so on - help you try to get some sleep (it will be hard!)
- Night dress or pajamas
- Laptop (if the hospital has free wi-fi!)

**Going in longer term (eg a resection or reconstruction surgery)**
Same as going in for a night plus
- Several pairs of underwear
- More extensive wash (toiletries) bag - shampoo, conditioner
- Yoga pants/loose trousers
- V-neck loose t-shirts
- Hair ties or clips to keep hair out of eyes
- Fleece blanket
- Load your iPad with movies and music - bring your headphones
- Ear plugs and sleep mask
- Bose Noise Cancelling ear buds – can be used to listen to movies and music, as well as just turning them on at night and popping them in your ears on silent as very effective ear plugs – highly recommended!
- Magazines (you can ask visitors to bring these too!)
- Any hobbies you can do while in bed (crosswords, knitting, crochet, drawing etc)
- A travel pillow to help support your head --- this was especially helpful for resection patients with the vampire stitches
- Sore muscle rub/balm. eg Icy/Hot.
- Extension lead with multiple power points for plugging in your laptop/iPad/phone/etc
- Netflix subscription! Great to watch a whole TV series while recovering
- Travel hair dryer - luxury!
Tips on surviving major surgery in hospital

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

Pre op be as healthy as possible. Lose weight if you have excess, eat really healthily, do as much exercise as you can. I definitely credit my overall recovery to this regime. I did the 5:2 diet for a year before my op - this reduces your sugar levels, bad cholesterol and puts your body into repair mode as well as weight loss. I am sure my 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. I never lost sight of what I wanted from my operation.

Try to recognize progress every day, however small it is. It will motivate and encourage you - for me it was little things - being allowed to use the bathroom rather than a bedpan, getting my feeding tube removed, being allowed to wash myself, then shower...even taking fewer painkillers was progress...

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

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

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

Talk - not talking is quite disturbing especially if you can't. People treat you differently and try to assume what you're thinking or trying to communicate. Work out the best way to communicate. I'm a touch typist so a Bluetooth keyboard with my iPad worked well - I'd have the iPad on view to my guest and type away all the things I'm feeling and thinking. The keyboard I had on my lap, beaming my words across to the pad!

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.
I cannot stress the value of having some noise cancelling earphones. Mine were tiny Bose in ear ones and they blocked out 80% of all sound even without playing music, just with the power on. I wore them every night. If someone wants to buy you a 'good luck in your operation' present this is what you want!

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

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

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

**Share** your experience with others who understand - namely the people in the Living with Idiopathic Subglottic Stenosis support group. The support and positive messages I received from everyone while I went through my recovery were so uplifting - definitely a boost to how I felt and coped throughout.

Finally **be positive.** I cannot stress the power of positive thought - think yourself well, laugh, smile and focus on the good rather than the bad.