

Nose Deformity And Reconstruction In The Individual With Systemic Vasculitis

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Nose Anatomy And Nose Reconstruction

The anatomy of the nose is divided into two distinct parts - the sinuses and the external nose. This article will mainly focus on the external nose and how it is affected in vasculitis and how it can be reconstructed.

The Anatomy

The external nose is a complex three-D structure. It is made of three layers. From outside inwards skin cartilage/bone framework and inner lining. Each of these 3 layers serves a purpose. We use the analogy of a house to explain the layered structure of the nose.

Skin

The skin serves as a protective barrier to the underlying structural layer. The skin envelope has various thicknesses between individuals, some have thin skin others have thick skin. The skin can be viewed as the cladding or paint that covers the house. If the cladding is thick the underlying structures, the bricks, (cartilages and bone) will be less pronounced. If the cladding is thin the underlying structures will be very visible and small irregularities will be visible to the eye.

Cartilage and Bone Framework

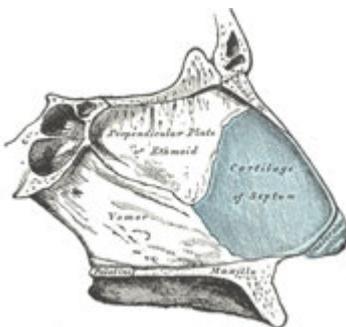
The framework of the house is similar to the bricks of a building. There are three main components, each one supporting a section (upper, middle and lower) portions of the nose.



The upper portion of the nose is supported by the **bones of the maxilla (cheek bone and nasal bones)**.

The middle third of the nose is supported by the **septum attached to the upper lateral cartilages**. These cartilages support the middle portion of the nose. Loss of support here will lead to a saddle nose deformity or in some cases a septal perforation (hole in the septum).

The lower third of the nose is supported by the **lower lateral cartilages**. These cartilages are curved and angulated outwards to give the characteristic shape to the tip and part of the sides of the nostrils (alar) and help support the tip and the nostrils (alar) of the nose.



The septum is attached to the upper lateral cartilages. The lower lateral cartilages and the nasal bones give the shape of the nose (modified to a degree by the thickness of the skin).

Inner Lining

The inner lining of the nose is made up of skin and mucosa. It is a vital layer of the nose and can be viewed as the foundation of the house. Any deficiencies (tears or gaps) allow bacteria normally carried in the nose, to infect the middle framework layer of the nose. This can cause damage to the cartilage of the nose and weaken the support structure. In addition when these gaps heal, they heal

with scarring and contraction that causes the nose, which is mobile, to become twisted producing an external nose deformity. The nostrils can become narrowed which can impair breathing. This is analogous to faulty foundation in a house leading to subsidence.

The Airway

The narrowest point in the whole respiratory tract is the internal nasal valve area, 1 cm inside the nostril. This is very important as small deflections or distortions of the nose can produce dramatic reductions in airflow and nasal blockage.

The Vasculitic Nose

Wegener's Disease - Granulomatosis with Polyangiitis (GPA)

GPA commonly affects the nose, it is the classical disease for nasal manifestations of vasculitis disease. The vasculitic diseases such as Wegener's (GPA) causes an inflammation of the medium and small sized blood vessels. When this occurs in the nose this causes inflammation in the mucosa. This leads to a continuous cycle of inflammation and healing with granulation (healing) tissue. The granulation tissue results in scar formation and tissue contracture that can progressively distort the nose. This process can be modified by the bacteria you contain within your nose amplifying this process.

Mucosal bleeding, nasal stuffiness and crusting in the nose and occasionally nasal pain are all signs of disease activity and should initiate a medical review from your Rheumatologist. Endoscopic examination of your nose, and blood test such as CRP, cANCA, PR3 MPO, are useful in monitoring disease activity. As this process continues untreated a stepwise progressive injury to the nose can occur.



Although most affected individuals have nasal symptoms, only some have progression to the more severe forms of nose deformity.

If the cycle of inflammation and healing continues, the cartilage in the septum is weakened secondarily to the inflammatory process. This can lead to a septal perforation (hole). As this perforation (hole) enlarges (see photo below), it can cause weakening of the support of the middle portion of the nose and can result in a saddle nose deformity in your nose. According to one large study the frequency of a nasal septal perforation was 33% and 28% for saddle nose deformity.



Septal Perforation

This is a defect in the mucosa and cartilage causing a hole in the partition of the nose

Severe Saddle

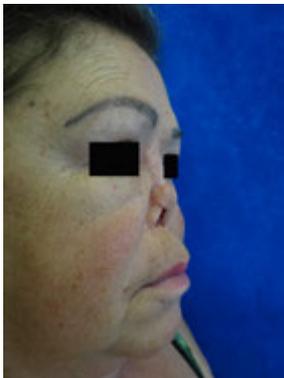


As the septum is progressively damaged the saddle nose deformity worsens and the nose becomes shortened and the tip of the nose rotates upwards.

This saddle deformity can be reconstructed in a single surgical procedure. Surgical intervention requires reasonably good general health and that the affected individual also be in long term remission.

With further damage from a cycle of inflammation and healing, the inner lining of the nose begins to shrink through scarring and further shortening of the nose can occur.

Severe Saddle And Shortening Of The Nose



In a minority of affected individuals this inflammation can extend full thickness (all three layers of the nose) as in this case. These cases are less common but they can result in contracture and shortening of the skin with or without a hole (fistula) between the skin and the nose (nasal cavity). These individuals will then have deficiencies in all three layers of the nose (skin, cartilage, lining).

(https://en.wikipedia.org/wiki/Granulomatosis_with_polyangiitis)

Some of the more severe deformities can be reconstructed. There are reports of correction of severe saddle and shortening of the nose. Some authors suggest rib grafting only, some authors also correcting the lining deficiency with flaps. There are also reports of closure of nasofacial fistula. These reconstructions can require several operations (3 to 4) each staged 3 weeks apart. It also requires reasonably good general health and a period of well established long term remission.

Other Types of Systemic Vasculitis

Relapsing Polychondritis – Has a variable affect on the nose. Various sub-sites can be affected so the nose can be spared. If the nose is affected it can mirror Wegener's disease with pain and nasal stuffiness progressing to perforation and saddle nose deformity

Systemic Lupus – rarely causes septal perforation < 5%, or nasal collapse/saddle.

Microscopic Polyangiitis – Often limited to crusting and epistaxis and sinusitis, progression to progressive nasal collapse is rare

Why Is The Nose Deformity So Debilitating?

Functional Concerns

Nasal obstruction can cause significant reduction in quality of life. It can cause difficulty with exercising, reduced quality of sleep and daytime fatigue. Not being able to breathe properly can affect your productivity and many aspects of your life.

Appearance Related Concerns

We all know that the nose is fundamental to the individual's appearance but why is this? What makes small changes in the nose result in dramatic changes to a person's overall facial appearance.

There are several reasons for this but a very important one is that we focus on the central face for facial recognition. We regularly discard much of the face when we try to recognize someone. So it follows that small changes in these key areas may affect how we recognize someone. A study, which performed 'Heat Maps' of how people visually scanned and recognized human faces. It showed what areas of the face we look at to recognize someone. It is fascinating that the eye mainly tracks the central face (the nose and the eye's). It is understandable therefore that nose deformity can be debilitating for some patient's identity.

Nose Reconstruction

Timing of Nose Reconstruction

Generally two years of remission is required before undertaking reconstructive surgery. However when one wishes to correct a more severe deformity, a longer period of remission is sometimes preferred.

Concepts In Nose Reconstruction

Nose deformity requiring reconstruction results from deficiency or (less common) excess in at least one of these 3 layers:

- Skin
- Framework
- Inner lining

In most instances nasal deformity results from a deficiency caused by cancer (skin or sinus cancer) or an inflammatory process such as vasculitis, that destroys the nose. In order to reconstruct the nose correctly we must correctly diagnose what is in excess or what is deficient. Each of these excesses or deficiencies in each layer must then be individually addressed and returned to normal size and strength in order to reconstruct the nose correctly. There are, unfortunately, no short cuts.

Skin

The least complex layer to reconstruct is the cladding of the house. So resurfacing a nose with skin borrowed from the forehead or cheek is not generally considered a complex procedure.

Framework

The next level of complexity is reconstructing the bricks of the house (the cartilage and bone framework). This requires harvesting a small portion of your rib or ear cartilage. This is more complex than re-cladding of a house and requires careful sculpting of these tissues. We only ever use your own tissue (autologous tissue), mainly from your ribs or ears. Although we use cartilage/bone from these areas the shape of the ribs or ears is unaffected.

Inner lining

The inner layer of the nose is called the inner lining. It gives no shape to the external nose and its function is simply to act as a barrier to bacteria present in your nose and to provide a blood supply to the framework layer from the inner aspect of the nose. It however is the most complex layer to reconstruct. Defects in the inner lining are analogous to defects in the foundations of a house. You will appreciate that faulty foundations in a house will lead to progressive subsidence and instability of the whole structure upon which it is built, and they are of course complex to correct.

Septal Perforation or (hole in the septum)

Septal perforations can occur from: previous septal surgery, vasculitic diseases, manual nose picking, cocaine use and 50% are idiopathic – or unknown cause

Perforations can cause little or significant functional symptoms for affected individuals. Small perforations can whistle when the affected individual breathes through their nose. Large perforations do not tend to cause whistling, they however can cause a feeling of being blocked and nasal crusting which can be very troublesome for the individual. In Vasculitic disease, nasal crusts and a sensation of blockage can also occur from the disease process itself even without a septal perforation. The mainstay of treatment is treating the underlying active disease, and any co-existing sinus disease in the acute phase. This is achievable with nasal saline sinus rinse and oral antibiotics if required (directed by your ENT Surgeon) and treatment of the underlying vasculitic disorder (directed by your Rheumatologist).

In the presence of vasculitis most septal perforations are not repaired. Lubrication with Vaseline and saline douches can help treat perforation related symptoms. When the disease is stable a bespoke septal button can be made from a CT scan to fill/obturate the hole. This is successful in some cases. Small septal perforations in symptomatic individuals who are in long standing remission could be considered for repair on a case-by-case basis, but this is not routinely offered.

Saddle Nose Deformity And Its Correction

Saddle nose deformity is not a disease, it is the clinical description/appearance of the nose that occurs following loss of support of the nose from destruction of the nasal septum. This lack of support produces a typical concavity to the bridge of the nose in side (profile) view – like the shape of a horse's saddle, hence the term 'saddle nose deformity'. This deformity progressively causes increased breathing difficulties and the appearance is quite distressing for many of the affected individuals. Any process that weakens the septum can produce a saddle nose deformity:

- Over resection/removal of the septum from poorly performed septoplasty, rhinoplasty
- Cocaine nose damage to the septum
- Vasculitic disease that weakens the septum through inflammation
- Large perforations in the nasal septum



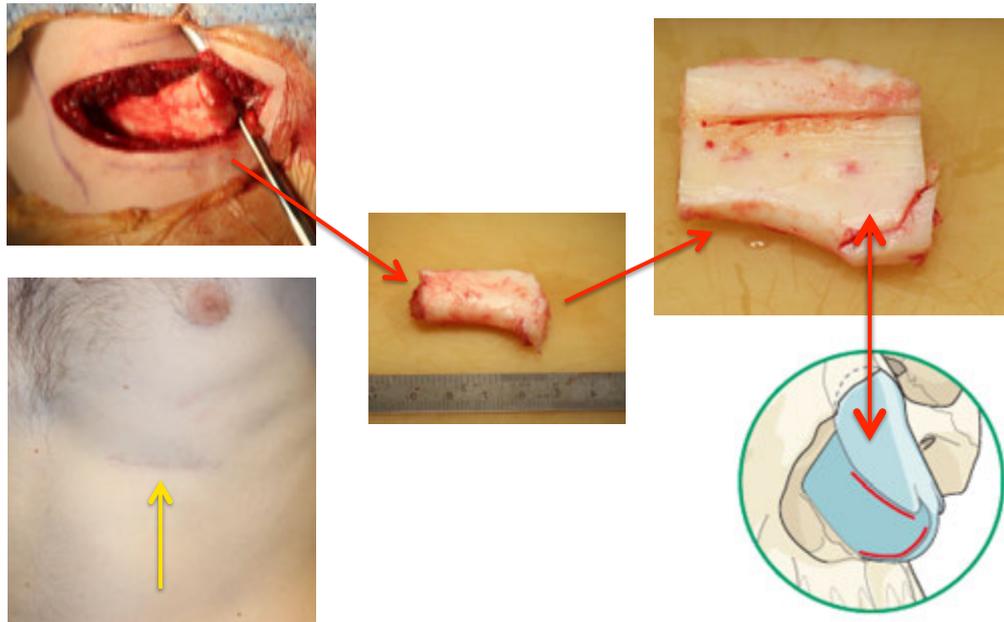
Correcting the saddle requires rebuilding of the septum to re-support the mid portion of the nose. this is done by taking cartilage grafts from ears or from your rib cartilage.

There are various grading systems to saddle nose deformity. The majority can be corrected in a one-stage procedure as a day case patient. Extreme cases may require more than one stage to successfully reconstruct the nose.



This case example (left) demonstrates nasal collapse, which has produced a severe septal saddle. The nose deformity is reconstructed with rib cartilage in one operation.

If necessary rib cartilage can be taken and sculpted to completely replace all of the septum as shown below. Yellow arrow indicates healed scar, in women this scar is hidden under the breast crease (without changing the shape of the breasts)



Can you avoid the use of Rib and Ear Cartilage – Synthetic Permanent Implants

The use permanent synthetic implants such as polyethylene, polytetrafluoroethylene (PTFE) or Silastic in vasculitic nose reconstructions is controversial. Implants are an attractive option as they avoid making a second incision in the ear or chest wall. It also great simplifies the surgery which why some surgeons like to use them. To date no group has published sufficient numbers of synthetic implant in vasculitic noses to safely recommend their use.

The main concern with the use of any permanent synthetic implant is that they expose the nose to a permanent continued lifetime risk of infection and extrusion (the implant coming through the skin of the nose). If this occurs the implant must be removed in all cases, and the reconstruction revised. This can occur at any time sometime up to 15 even 20 years after surgery. If implant extrusion occurs in a reconstructed nose it is a complex problem to correct. This stands true for normal noses, whereas in vasculitic noses the chances of infection and extrusions are almost certainly higher. It is our opinion that the use of any permanent synthetic implant in nose reconstruction is not justified. Out of a survey of 5 leading international nose reconstructive surgeons, none recommended the use of permanent implants.

Nasal Full Thickness Reconstruction

When conditions affect the full thickness of the nose there can be deficiencies in all 3 layers of the nose. This is rare in vasculitis but more common in cancer reconstruction. Using case studies from nose reconstruction shown below we demonstrate how the nose can be reliably rebuilt even if the majority of the nose is missing.

Half Nose Missing From Cancer



Three layers of the nose are reconstructed. Using techniques from skin cancer nose reconstruction a normal nose can routinely be achieved over 3 operations, 3 weeks apart. Only the individual's own tissue is used with no implants to reduce this risk of infection. The patient now has a functioning nose that appears normal at conversational distance (however faint scars are still noticeable).

Majority of Nose Missing From Cancer



Three layers of the nose are reconstructed. Again using techniques from skin cancer nose reconstruction a normal nose can be achieved over 4 operations, each 3 weeks apart. Only the individual's own tissue is used with no implants to reduce the risk of infection. The patient now has a functioning nose that appears normal at conversational distance (however faint scars are still noticeable).

The standard for nose reconstruction for cancer is a symmetrical nose with no nasal obstruction and only faint visible scars on the nose. In vasculitis however the rate of infection and graft absorption in nose reconstruction is likely to be higher than in normal noses. This is due to the fact that the grafts and flaps are placed in a relatively hostile environment, as the remaining skin of the nose can be scarred with a relatively poor blood supply. This makes surgery more demanding. Despite this, nose reconstruction in vasculitic disease may be beneficial and a worthwhile endeavor. Some Surgeons nevertheless feel the risks associated with complex reconstruction are still too high for it to be offered routinely. Therefore as an alternative to surgery in certain cases prosthetic rehabilitation is well tolerated and impressive results can be obtained in expert hands.

Prosthetic Rehabilitation

Nasal prosthesis can be fashioned to correct complex deformities. This can either be as a temporary measure till surgery can be scheduled or as a permanent management for the patients who wish to avoid surgery or patients medically unfit to undergo several surgeries..

Although the prosthesis can avoid multiple operations (3-4) associated with surgical correction of severe deformity they do have several issues.

Retention of the prosthesis can cause difficulties. They are usually initially retained with adhesive glue which can cause skin reactions. In addition skin glue can sometimes fail with resulting detachment of the prosthesis. This is naturally a concern for many prosthetic patients. It is for these reasons that patients with long term prosthesis will often opt for osseo-integrated implants to retain the prosthesis. A surgical procedure is required to place these titanium implants around the nose and they are then left to integrate with the bone over 3-6 months, which are successful in 80-90% of cases. Following this magnets are placed onto the integrated implants. 2 -3 magnetic implants are required to retain one nose prosthesis. One other main issue with implants is the continual need to attend hospital as these latex nose prosthesis degrade over several months and in some cases last only 1-2 months. Therefore replacement prosthesis and repeat fitting sessions are required. The cost implication of this is significant at approximately 1000 pounds per year (UK NHS figures) if the patient is not covered by health insurance or the National Health Service.

It is for these reasons that decisions to proceed with surgical reconstruction of complex nasal defects or opt for prosthetic rehabilitation in vasculitic patients are difficult to make and should be individualized for each patient.

The Failed Nose Reconstruction

All surgeons have complications and occasionally suboptimal results. If a patient remains unhappy with the appearance or has continued nasal obstruction following a nose reconstruction they should seek an opinion regarding revision surgery. If the original operating surgeon is not happy to do so, they should seek a second opinion. It has been our experience that the information previously provided to patients with failed nose reconstructions that 'nothing more can be done for you' may not always be correct.

Summary

Nose reconstruction is a highly specialized area of surgery with few Surgeons undertaking complex reconstructions. When this is combined with a rare disease such as vasculitis it then becomes understandable that there is still a degree of uncertainty in some areas of its management. A body of evidence already exists demonstrating that it is generally safe to correct moderate (saddle) nasal deformity when in long-term remission, and it should be offered routinely to patients if their medical condition allows reconstructive surgery. This body of evidence does not yet exist to make the complex reconstructions routine however it may in time, as experience grows. Till that time personalized decision making is key.

Is Nose Reconstruction Funded On The NHS?

Nose reconstruction for vasculitis disease is funded on the NHS, therefore affected individuals who are eligible for NHS treatment, do not need to pay for their surgery.

The Future

Our group in conjunction with Department Of Tissue Engineering At Nottingham University and other groups around the world are working on a tissue-engineered nose grown from the affected individual's own stem cells. We hope that through this research we can move to repair a whole nose in a reduced time. From more information on our ongoing research on nose reconstruction you can visit our website: <http://www.nosereconstruction.co.uk/research/>

Key points for the individual with vasculitis and nasal deformity:

Vasculitic disease can affect the sinuses and external nose

Degree to which it does depends on the type of vasculitis you have

Wegener's is the most common entity to cause external nose deformity and the degree to which it does this is variable

Septal Perforations are not classically repaired in the setting of vasculitis

Saddle nose deformities can be repaired in remission

Combined nasal and facial defects can be repaired in remission if the affected individuals general health allows several surgical procedures

The use of any permanent nasal implant in the reconstruction of a vasculitic nose, should in our opinion be avoided

Complex nose reconstruction is highly specialized area of surgery

Nose reconstruction surgery for vasculitis is currently funded on the NHS

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