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Avoiding pitfalls in open augmentation rhinoplasty with autologous L-shaped costal cartilage strut grafts for saddle nose collapse due to autoimmune disease: The Cambridge experience☆

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Summary *Introduction:* Saddle nose deformity due to autoimmune diseases such as Wegener's Granulomatosis and Relapsing Polychondritis is aesthetically, functionally and psychologically distressing for patients. However, "reliable" options for surgical correction remain limited in the literature. We present our experience of augmentation rhinoplasty in this patient population focussing on the techniques and pitfalls of L-shaped costal cartilage grafting.
Methods: Five patients undergoing rhinoplasty for saddle nose deformity due to an autoimmune condition were identified over an 11-year period at a major tertiary centre. All patients were in remission from their condition at surgery and underwent L-shaped costal cartilage grafting at augmentation rhinoplasty. Case notes were reviewed retrospectively.
Results: All patients achieved a marked improvement in nasal position, shape and contour and were very pleased with their overall appearance. The average length of follow up was 2.8 years. There were no infections, graft exposure or warping. No resorption of cartilage was observed and there have been no recurrent deformities.

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Conclusion: This case series describes a possible approach to corrective rhinoplasty in patients with saddle nose deformity caused by autoimmune disease, highlighting the key technical steps and potential pitfalls of intraoperative and perioperative care in this population. The approach is straightforward, reproducible, and achieved pleasing aesthetic outcomes and high patient satisfaction. Given careful planning and meticulous execution, L-strut cartilage grafts for augmentation rhinoplasty to correct saddle nose deformity in these patients is of great benefit.

Level of Evidence: Therapeutic Study Level IV, case series with pre/post test.

Introduction

The surgical treatment of saddle nose deformity resulting from autoimmune conditions such as Wegener's Granulomatosis (WG) and Relapsing Polychondritis (RPC) is technically challenging. WG is an idiopathic necrotizing vasculitis with an incidence of 50—100 per million population in Europe and predominately affects the upper respiratory system, kidneys and lungs.[1] Treatment for the condition has advanced remarkably in recent years and current medical management with corticosteroids, immunosuppressants and novel biological agents are successful in limiting disease activity with remission achieved in 90% of cases.[2] However, in cases where therapy has not been optimised, serious complications such as saddle nose deformity can result.[3]

Saddle nose deformity occurs in 10—25% of patients with WG.[1] Sino-nasal manifestations are common and only second to the lung as a site for presenting symptoms (67% of all patients) and eventual involvement (91%).[4] The nose is vulnerable to damage as it is often the first area to be affected and is prone to disease relapse.[2] Inflammation of the vessel walls causes rhinitis and nasal crusting, leaving the tissue friable and ulcerated. This can lead to nasal septal perforation, loss of structure and consequent saddle nose deformity. This deformity has both aesthetic and functional complications, with blockage of the nasal airway and reduction in exercise tolerance, and is associated with both physical and psychological morbidity.[5]

RPC is an immune-mediated condition predominantly involving the ears, nose, joints and respiratory tract that leads to the degeneration of cartilage and subsequent fibrosis. This can also lead to severe saddle nose deformity. The main stay of treatment involves immunosuppression with steroids and steroid sparing medications.

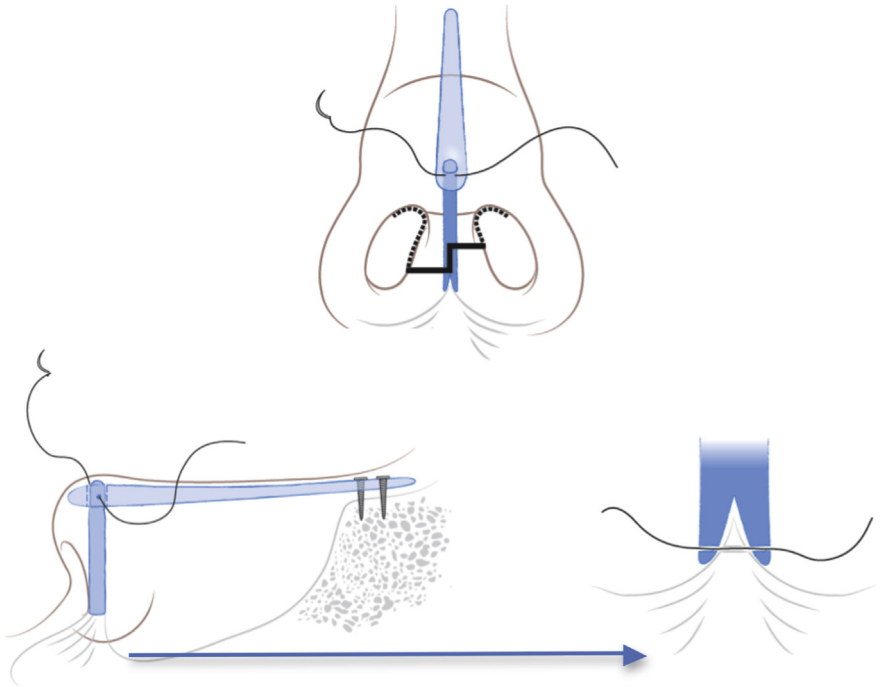


Figure 1 Artist's illustration showing the key points in the open rhinoplasty approach including the columellar and infracartilaginous incisions, the use of titanium monocortical screws to secure the dorsal strut to the nasal bone and the distal splitting of the columellar strut prior to fixation to the nasal spine.

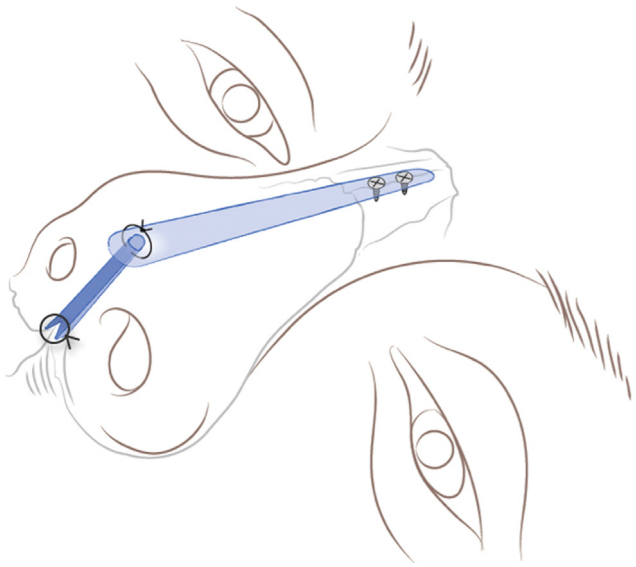


Figure 2 Artist's illustration demonstrating the completed open augmentation rhinoplasty with L shaped costal cartilage struts.

While the individual pathophysiologies of WG and RPC are different, both lead to almost identical cosmetic deformities and for this reason have been considered together in this case series.

The treatment of saddle-nose deformity and its associated symptoms falls into two categories: symptomatic and definitive. Conservatively, treatment for nasal crusting can be initiated with nasal saline rinses, topical steroids, mucolytics, and emollients which may be complemented with minor surgical operations including polypectomy and mucosal-sparing techniques.[1] The only definitive treatment is surgical correction via nasal reconstruction.[3]

Unfortunately, there are few reports in the literature exploring the correction of saddle nose deformity in patients with WG or RPC.[2,4,5,7,8] In this case series, we present our experience of open augmentation rhinoplasty using L-shaped costal cartilage grafts in patients with autoimmune disease and highlight key intra-operative and peri-operative features to optimise outcomes.

Patients and methods

Patient population

All patients undergoing correction of saddle nose deformity due to an autoimmune condition between 2008 and 2011 by a single consultant plastic surgeon with a specialist interest in cosmetic surgery (CMM) were identified from the theatre records, consultant's log book and electronic billing records. Surgery was performed at a tertiary university teaching hospital or the Cambridge Private Hospitals. Patients were followed up in the outpatient clinic for a minimum of one year from surgery.

Case notes were reviewed retrospectively, focussing on demographics, severity of autoimmune illness and deformity, operative detail, complications and cosmetic outcomes.

Surgical technique

All operations were performed under general anaesthesia. An open rhinoplasty approach was used in all cases comprising a stair-step transcolumellar incision extended into bilateral infracartilaginous (rim) incisions (Figure 1). The lower lateral cartilages were then carefully exposed, and dorsal dissection of an extra-mucosal pocket to the radix was performed to expose the underlying nasal bones and cartilage remnants. An additional 1 cm skin crease incision over the radix was made to allow direct access to the nasal bone cranially. The dorsal cartilaginous structures were examined for extent of disease damage. The dorsum of the nasal bone and residual dorsal cartilage were then rasped and trimmed, as necessary, to create a receptive graft bed.

Costal cartilage was harvested from the sixth or seventh rib via an inframammary skin incision followed by electrocautery dissection through the fascia and pectoralis major and rectus muscles. Following subperichondrial costochondral harvest, a water test via the valsalva manoeuvre was performed to assess for any pleural leaks.

Two pieces of cartilage were then fashioned - the dorsal strut (length 3.5—5.0 cm; variable width) and the

Table 1 Patient Characteristics

Patient	Gender	Age	Saddle nose severity	Autoimmune disease	Comorbidities	Immunosuppression at surgery	Follow up*
1	F	25	IV	WG	None	Prednisolone 10mg daily Budesonide nebulisers 1mg BD	5.05 years
2	F	74	IV	WG	Spinal degeneration Hypertension Depression	Prednisolone 1.5mg/ day Methotrexate 22.5mg/ week	3.01 years
3	F	32	IV	RPC	None	None	2.68 years
4	M	30	III/IV	WG	Crohn's disease	Prednisolone 10mg/day Azathioprine 150mg/day Mesalazine 1g/ day	1.57 years
5	F	30	III	WG	Depression	Prednisolone 1mg/day Azathioprine 100mg/ day	1.52 years

Key WG = Wegener's Granulomatosis, RPC = Relapsing Polychondritis
* From date of operation to article submission

Figure 3 (Patient 1 in Table 1) © Addenbrooke's Teaching Hospital.

- A 25 year old female with type IV saddle-nose deformity secondary to Wegener's Granulomatosis and previous failed attempts at dorsal augmentation with alloplastic material.
- At 2 years following revisional surgery, please note the maintenance of excellent cosmetic results. The columellar retraction, dorsal depression and splaying have been corrected, there is no evidence of resorption (a—d).

columellar strut (length 2.5—3.5 cm; width 5 mm, height 2—3 mm). The cartilage was carefully carved using the balanced forces concept of Gibson and Davis with a focus on symmetry to reduce the likelihood of post-operative warping.[9] The dorsal graft was then inserted into the dorsal pocket and, once manoeuvred into the correct position, secured with two titanium self-tapping monocortical screws (6 mm—8 mm length) to the nasal bone to ensure a stable and symmetrical reconstruction (Figure 1).

One end of the columellar graft was carefully split for approximately 5 mm to allow it to sit astride the nasal spine/crest. This was fixed to the nasal spine with a 4/0 PDS suture through a hole drilled in the spine with a 1.5 mm dental drill (Leibinger Wurzburg Miniplating System, Wurzburg Germany) (Figure 1). The two grafts were then connected with each other via a hole and dowel arrangement and fixed with a single 4/0 PDS suture to ensure a secure join (Figures 1 and 2). The overall shape was then examined and the cartilage further trimmed to improve contour as needed.

After securing the L-shaped graft, the chest wall donor site was closed in layers using 2/0 PDS to the muscle fascia and 3/0 Monocryl to the deep dermal and subcuticular layers. The rim columellar incision was closed with 4/0 Vicryl Rapide to the mucosa and 5/0 nylon to the skin. The radix incision was closed in two layers (5/0 Vicryl and 6/0 Nylon). Half inch skin tapes (3M™ Steri-Strip) and a thermoplastic external nasal splint were applied and remained in place for 7 days.

Results

From 2008 to 2011, five patients (four female, one male) underwent open rhinoplasty with L-shaped rib cartilage grafts for saddle nose deformity secondary to autoimmune disease (Table 1). Their mean age was 38.2 years (range 25—74). All patients had severe saddle nose deformity of at least Daniel and Brenner Types III and IV[10] secondary to either Wegener's Granulomatosis (WG, four patients) or Relapsing Polychondritis (RPC, one patient). At the time of surgery, all patients were in remission from their autoimmune disease. Four patients underwent a primary rhinoplasty, while one patient was referred as a secondary rhinoplasty to revise a prosthetic nasal reconstruction that had previously failed due to infection.

In terms of cosmetic outcomes, all patients achieved a marked improvement in nasal position, shape and contour following surgery and were very pleased with the improvement in appearance (Figure 3—5). In patient 3, some asymmetry of nostrils, length deficiency and left nostril bulge was noticed during follow up. This may have been due to 8 mm of the caudal part of the dorsal graft being inadvertently broken during handling after fixation to the nasal spine and insertion of columellar strut. This was surgically revised 15 months after the initial procedure. Other patients displayed mild imperfections not requiring further surgery — patient 1 and 4 (Figures 3 and 5) were noted to have minor dorsal deviation, patient 2 (Figure 4) had foreshortening and patient 5 reported asymmetry between nostrils. These patients elected not to have any reoperations as they were pleased with their overall

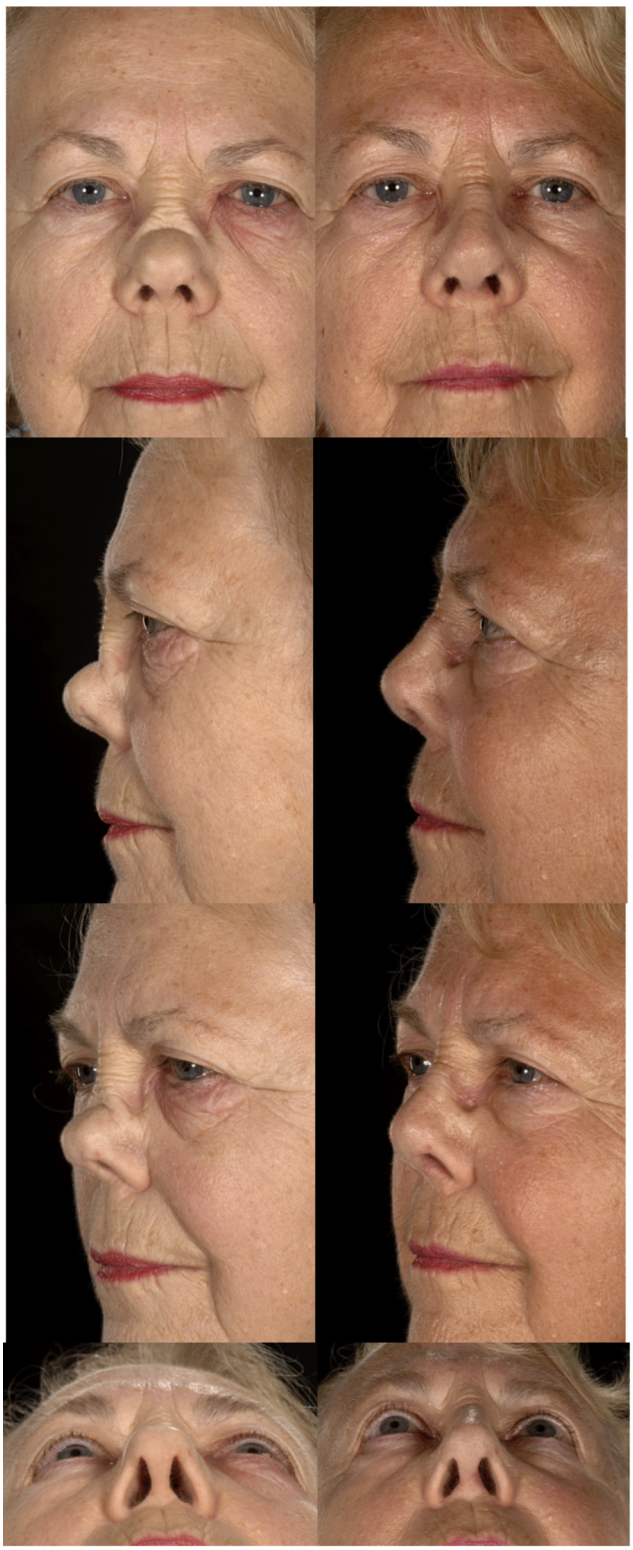


Figure 4 (Patient 2 in Table 1) © Addenbrooke's Teaching Hospital.

- A 74 year old female with a type IV saddle nose deformity secondary to Wegener's Granulomatosis.
- Please note the excellent post-operative appearance at 6 months with correction of dorsal depression and splaying (a—d).

cosmetic outcomes — especially with the degree of improvement.

Average length of follow up from the operation date was 2.8 years (range 1.52—5.05). Most patients did not experience a relapse in their autoimmune disease during this period. However, patient 5 experienced a mild flare in Wegener's symptoms two months following the surgery. This was controlled with the addition of rituximab to her immunosuppression regime and did not result in any nasal instability or further complications.

There were no other surgical complications including bleeding, pneumothorax, infection or poorly controlled pain. There have been no incidences of deviation of the nose due to warping. All donor sites healed satisfactorily.

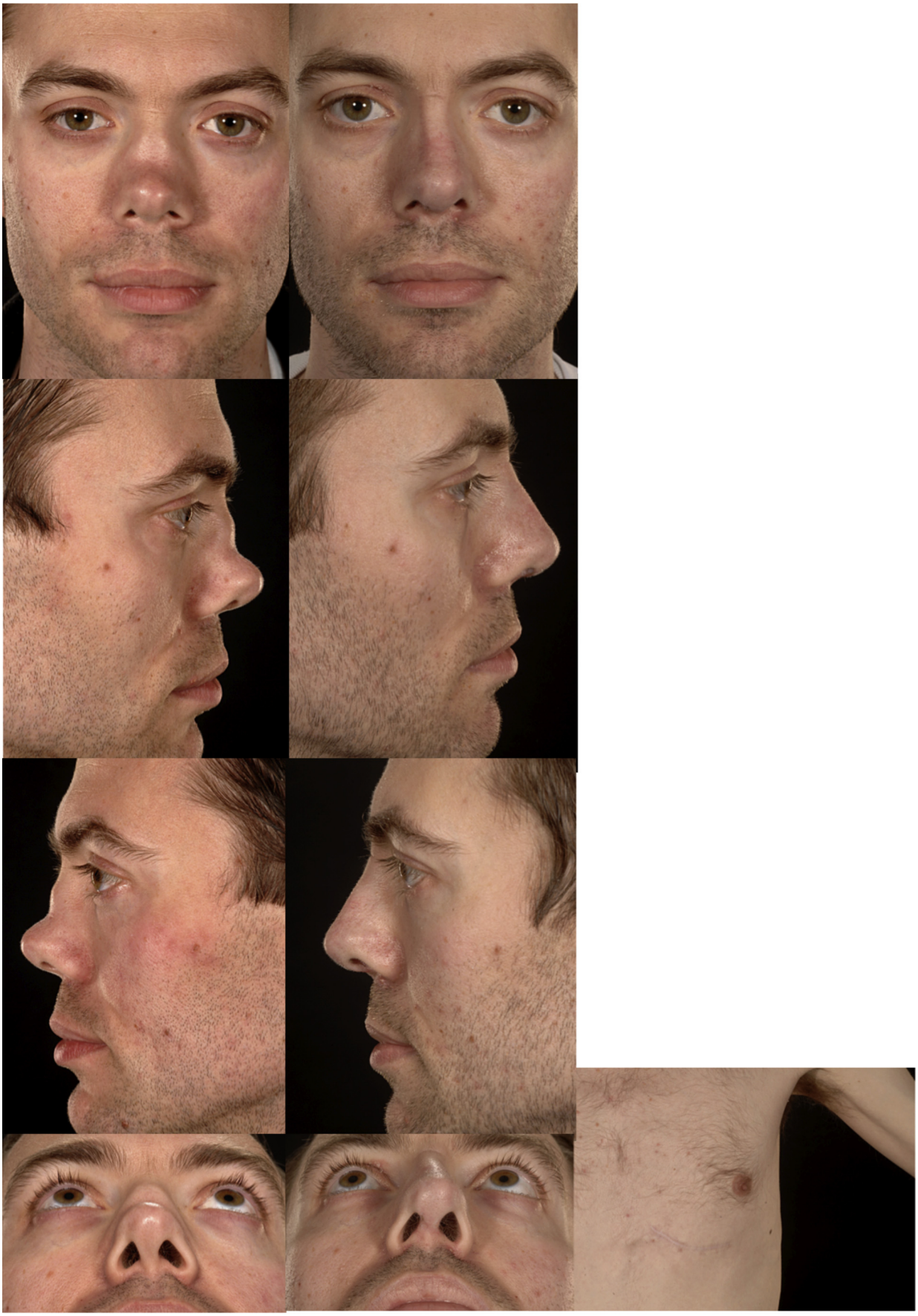
Discussion

In Wegener's Granulomatosis (WG) the repair of saddle-nose deformity has been shown to be a safe and effective procedure[1—3,5] with both functional and psychological benefit.[5] However, augmentation rhinoplasty continues to be underutilized in this population and only a limited number of case studies exploring rhinoplasty in WG have been reported.[2,4,5,7] In patients with Relapsing Polychondritis (RPC), surgical management for saddle nose is cautioned even in patients with quiescent disease due to disease recurrence.[11] In both WG and RPC more data regarding perioperative management and operative technique are required.

From our experience we have distilled the salient points important for optimising the chances for successful outcomes (Table 2). In addition to external cosmesis, the key anatomical considerations in planning an augmentation rhinoplasty for saddle nose deformity due to inflammatory causes (such as WG and RPC) include the height and projection of the nose, external skin defects, internal nasal lining defects and obstruction of nasal airways. Structural deficits may be large, with extensive septal defects and perforations between the two nostrils in place of where the cartilaginous septum used to be. In these cases, it is important to ensure, on clinical examination, that there is sufficient residual nasal mucosa to cover the grafts both in the columella and dorsum of the nose to reduce post-operative intranasal drying and crusting.[4] If there is insufficient nasal mucosa, other surgical options including flap reconstruction may need to be considered.[4]

Although a range of graft materials has been explored in augmentation rhinoplasty, the gold standard in saddle-nose deformity due to WG is costal cartilage.[7] In WG, costal cartilage is recommended as cartilage from other regions (i.e. auricular) are insufficient for large septal defects.[12,13] In RPC, bone has been the chosen grafting material as it is believed the presence of autoimmune chondritis precluded cartilage grafting due to a high risk of subsequent cartilage destruction.[8] For our patients, we chose autologous rib cartilage grafts for their pliability, strength and ready

- Some foreshortening remained, however the patient was pleased with the improvement and elected not to re-operate (b and c).



availability to meet the demands of large defects. We felt that this was also a reasonable option in our RPC patient as she had been in remission with no active steroid or immunosuppressive treatment for seven years prior to surgery. Furthermore, at surgery no involvement of the costal cartilage was noted and post-operatively there was no disease activity to suggest cartilage graft compromise.

The risk of postoperative warping of cartilage grafts is reduced when the struts are carved symmetrically according to the principles established by Gibson and Davis.[9,14] In our experience with strict adherence to this principle, warping has not been an issue, with all patients satisfied with the aesthetic result as compared to the pre-surgical appearance. We prefer to use costal cartilage from the 6th rib, an area that is thought to behave similarly to septal cartilage and is adequately pliable for carving and moulding.[15]

We choose to close the donor site after the cartilage grafts have been carved and secured in place at the graft site to enable the re-harvest of further cartilage in cases of mishap during the shaping, inset and fixation of cartilaginous struts. This also allows for left-over cartilage to be replaced, reducing the risk of chest wall deformity at the donor site.

Grafting method in augmentation rhinoplasty depends largely on the severity of the defect to be corrected.[7] In our patient population, we found the use of L-shaped strut grafts comprising separate dorsal and columellar components provided excellent support and structure. In order to prevent the dorsal component from breaking away from the columellar strut, they are fashioned in a dowel and hole arrangement secured with a holding PDS stitch.

Solid fixation of the L-strut to the graft site is key to maintaining the long-term structural stability of the rhinoplasty. While traditionally the graft has been secured with sutures at the caudal end only,[7,16] we have chosen to fix the graft at both cranial and caudal ends with a combination of suture and non-suture methods to minimize misalignment.

Microscrews, used in the fixation of cantilever nasal grafts in patients with extensive nasal defects, produce adequate compressive forces to ensure stability and rapid bone healing for good cosmetic and functional outcomes.[17–19] It is important to insert two screws to prevent graft rotation and provide adequate support to withstand significant tension from surrounding soft tissue.[20] Should one screw loosen, another remains in place to secure the graft. The screws must not protrude into the nasal cavity as exposure can lead to infections. No post-operative infection was reported in our case series.

In our practice, we prefer rigid fixation with two titanium self-tapping microscrews to secure the proximal end of our dorsal strut to the bony radix where possible, using X-ray to confirm their position. On one occasion, a single screw was used when the patient had had a long history of ENT procedures with insufficient nasal bone to support a second screw. However, the use of the double screw technique is generally reproducible and has led to good postoperative outcomes with no documented complications.

To prevent buckling or malposition of the columellar component, we first split its lower portion to enable it to sit securely on the anterior nasal spine, strengthening the join with sutures - one of which passes through the drill hole made in the nasal spine. A similar principle is applied to the join between the dorsal and columellar parts of the L strut.

While operative technique is crucial to the outcome of the augmentation rhinoplasty, perioperative factors must also be considered (Table 2). Due to the complexity of autoimmune disease, a multidisciplinary approach is recommended to ensure optimal care for patients during this period.

The timing of surgery must be considered in context of disease status and is best planned in consultation with rheumatologists. Valid concerns exist regarding the viability of rhinoplasty in patients with autoimmune inflammatory conditions as the underlying condition may cause an exaggerated inflammatory response to the graft.[21] Secondary infection is another serious risk as many patients are chronically colonised and have impaired host defences due to long term steroid and immunosuppressive therapies.[21] However, several case series have demonstrated positive outcomes in this patient population[2,5] and it is now recommended that rhinoplasty for saddle nose deformity in quiescent WG is safe and effective.[1–3,5] In RPC, surgery has traditionally only been recommended in cases of severe respiratory or cardiac complications, with no published data regarding rhinoplasty in patients with minimal disease activity.[6,8]

Quiescent disease at surgery is believed to result in positive surgical outcomes.[2] Following exacerbations of disease, surgery should only be considered after an appropriate waiting period. Caution is advised for patients on high doses of steroids as the increased risk of infection would be disastrous in a complex graft. To our knowledge, no specific perioperative guidelines exist regarding suitable waiting periods or 'safe' prednisolone doses in this group of patients. At our major tertiary centre, an informal protocol produced in collaboration with the rheumatology department recommends patients achieve at least six months of quiescent disease activity and are on a maximum acceptable dose of 10 mg Prednisolone daily prior to surgery. In our experience, this has resulted in excellent outcomes, few complications and no infections.

A thorough pre-operative assessment of the airway is an important consideration as obstruction (tracheal stenosis, subglottic stenosis) is a serious complication of disease involving the airways.[1,6] If there is any doubt, an anaesthetic or ear, nose throat specialist opinion should be sought. All of our patients had otorhinolaryngological assessments of their airways prior to referral.

Specific post-operative considerations in patients with WG or RPC are important in achieving good outcomes. To prevent infection, we routinely provided antibiotics, initially

Figure 5 (Patient 4 in Table 1) © Addenbrooke's Teaching Hospital 28/9/11 21/3/12 (6 months).

- A 30 year old male with Type III/IV saddle nose deformity secondary to Wegener's Granulomatosis.
- Postoperative appearance at 6 months (a—d).
- Note the excellent donor site scar in this male patient. This will be even better hidden in women due to larger breast tissue (e).

Table 2 Avoiding the pitfalls of augmentation rhinoplasty in autoimmune diseases

Pre-operative considerations	
Suitability for surgery	Disease activity
	- Minimal to no disease for at least 1 year
	- Maximum prednisolone dose 10mg/day
	Adequate mucosa to cover cartilage graft to avoid infection
	Adequate nasal bones to enable a secure graft join and prevent post-operative deformity
Pertinent referrals	ENT or anaesthetics: assess tracheal or subglottic stenosis
Intra-operative considerations	
Graft material	Costal cartilage: easier to carve, strong, adequate supply for large defects
Cartilage handling	Gibson and Davis' Principle of Balanced Forces to reduce warping[9]
	Careful handling of cartilage to avoid breakage
	Closure of donor site after securing graft
	- Allow for re-harvest in case of graft breakage
	- Replacement of excess cartilage to donor site to reduce deformity
Graft fixation	Joining dorsal graft to nasal bone
	- Minimise rotation Joining columellar graft to nasal spine
	- 2 x titanium self-tapping microscrews
	Joining dorsal and columellar components of L-strut
	- Hole drilled in dorsal graft, columellar graft inserted like a peg
	- Holding stitch
	Joining columellar graft to nasal spine
	- Split distal end of graft to sit securely on anterior nasal spine
	- Secure with suture drilled through nasal spine
Pain relief	Bupivicaine: intra-wound and subcutaneous at donor site
Post-operative considerations	
Wound care	Nasal pack: 1-2 days
	Nasal splint and sutures: 1 week
Infection prevention	IV Broad-spectrum antibioticsa: 24 hours post surgery
	Oral antibiotics: Following IV antibiotics, 5-day duration
	Topical antibiotic ointmentb: apply BD to columellar incision
Other precautions	IV Dexamethasone 8 - 12mg q8h for 24 hours: reduces postoperative swelling, nausea, vomiting

[a]i.e. Co-amoxiclav.
[b]i.e. Polymyxin and Bacitracin combination.

intravenously and then orally, in conjunction with a topical antibiotic cream (Polyfax: polymyxin and bacitracin) applied to the columellar and rim incisions. In addition, we made sure to provide adequate steroid cover via intravenous dexamethasone to reduce post-operative swelling and vomiting.

Obvious shortcomings of our case series are the small number of patients and relatively short follow-up period. However, our patients had a high level of satisfaction with the nasal shape and contour following surgery. Although some minor cosmetic issues of asymmetry remained in four out of five patients with one electing for a further revisional rhinoplasty, all patients were very pleased with the cosmetic and functional outcomes. All patients healed well and there were no major immediate or delayed complications from the surgery or the underlying autoimmune condition. This is consistent with other published success rates,[2,5] further validating our surgical approach in this patient population.

Conclusion

This case series describes a possible approach to augmentation rhinoplasty in patients with saddle nose deformity caused by autoimmune disease. This technique has resulted in pleasing aesthetic outcomes and high patient satisfaction. Importantly, the technique has proven to be reproducible and straightforward with consistent results over many years in the hands of a low-volume operator. The present report also outlines the pertinent precautions needed to achieve positive surgical outcomes in this challenging patient population. We firmly believe that, given sound surgical technique and thorough peri-operative planning, L-strut cartilage graft augmentation in the correction of saddle nose deformity in patients with Wegener's Granulomatosis and Relapsing Polychondritis can be of great physical, functional and psychological benefit.

Consent

The patients were fully consented for the surgery and provided both verbal and written consent for the use of the images and case histories in this article.

Conflicts of interest

The authors have no other financial interests, commercial associations or personal relationships to declare in relation to the contents of this article.

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