Wegener granulomatosis (WG) is a life-threatening autoimmune disease of unknown etiology first described in 1936. The classic clinical triad consists of necrotizing granulomatous inflammation of the respiratory tract, necrotizing glomerulonephritis, and systemic vasculitis. Prior to the introduction of modern immunosuppressants, WG was usually fatal due to renal complications. Involvement of the eye and adnexal structures is common, occurring in 29% of patients. Nasolacrimal duct obstruction (NLDO) following necrosis of the nasal tissue is seen in 7% of patients, who may seek relief through lacrimal surgery.

Dacryocystorhinostomy (DCR) has been shown to be a safe, effective intervention but failures and complications are more common in this patient group. While conventional teaching recommends an external approach, increasing use of the endonasal approach (EN-DCR), with or without an endoscope, raises the question of whether it can be used safely and successfully in WG. We present 4 cases treated endonasally (cases 1 and 2 using endoscopy by D.S., cases 3 and 4 using direct visualization by P.J.D., customary for each surgeon) and offer further advice on management of this patient group.

Report of Cases. Case 1. A 54-year-old woman, diagnosed 4 months previously with WG and treated with prednisolone and cyclophosphamide, presented with a 2-year history of right epiphora and recurrent episodes of acute dacryocystitis. Syringing and dacryocystogram showed complete NLDO. Computed tomographic scan demonstrated destruction of the ethmoids, medial wall of the left maxilla, and anterior cribiform plate with a narrow nasal cavity. Surgery was deferred until her disease was controlled and 20 months later endoscopic EN-DCR was performed, with Ears, Nose, and Throat assistance because of grossly disturbed anatomy. The lacrimal bone was eroded and there was dense scar tissue adjacent to the nasolacrimal sac; perioperative prednisone (1 mg/kg tapered over 4 weeks) was administered. The patient was asymptomatic following surgery and the ostium was patent at 14 months using the endoscopic dye test.

Case 2. A 49-year-old woman presented with a 12-month history of left epiphora and a preceding episode of acute dacryocystitis and orbital cellulitis. She had known WG for 2 years associated with severe sinusitis and a stricture in the left main bronchus and was treated with prednisolone and azathioprine. She had a saddle nose deformity due to destruction of the nasal bridge. Syringing showed complete NLDO and computed tomographic scan showed a deviated septum but good resolution of sinus disease. Left endoscopic EN-DCR and septoplasty were performed leading to relief of epiphora and a patent ostium on the endoscopic dye test at 12 months.

Case 3. A 47-year-old woman presented with 6 months of left epiphora complicated by 2 episodes of dacryocystitis, once requiring abscess drainage. She had a 10-year history of WG controlled with cyclophosphamide treatment; her last clinical recurrence had been 2 years previously and results of a recent antineutrophil cytoplasmic antibody test were negative. Left nonendoscopic EN-DCR was performed and she was symptom free and irrigated freely 6 months later. At that time, she had developed right NLDO and an EN-DCR was performed with Ears, Nose, and Throat assistance. The patient was asymptomatic following surgery and the ostium was patent at 14 months using the endoscopic dye test.

Case 4. A 47-year-old woman presented with 10 months of left epiphora following a preceding episode of acute dacryocystitis and orbital cellulitis. She had known WG for 2 years associated with severe sinusitis and a stricture in the left main bronchus and was treated with prednisolone and azathioprine. She had a saddle nose deformity due to destruction of the nasal bridge. Syringing showed complete NLDO and computed tomographic scan showed a deviated septum but good resolution of sinus disease. Left endoscopic EN-DCR and septoplasty were performed leading to relief of epiphora and a patent ostium on the endoscopic dye test at 12 months.

**Figure 1.** Case 1. A, Narrowing of the nose due to Wegener granulomatosis. B, Coronal computed tomographic scan showing destruction of the ethmoids and the right nasolacrimal duct causing obstruction.
formed on that side. A year later, she
devolved recurrent right epiphora
during a flare-up of her WG, with
granulation tissue obscuring the
original ostium. Once this was
controlled medically, a repeated
EN-DCR was performed and she has
been epiphora free for 5 years.

Case 4. A 49-year-old man pre-
sented with left optic neuropathy
and apical compression. His com-
puted tomographic scan showed a
left orbital apical mass lesion
(Figure 4) and WG was diag-
nosed from the biopsy specimen. Al-
though visual acuity returned with
systemic prednisolone and metho-
tretate treatment, he developed right
NLDO 3 months later. Once the dis-
ease was deemed to be quiescent 4
months later, a right nonendo-
scopic EN-DCR was performed with
insertion of Crawford tubes. These
were removed 3 months later and the
patient remained asymptomatic 9
months following surgery.

Comment. These cases demon-
strate the complexities of manag-
ing NLDO in patients with WG,
similar to previous case series,3
and that EN-DCR can be used
successfully in this group. How-
ever, various arguments have been
raised against EN-DCR in WG,
including disease reactivation,
ostium size, bleeding, and endo-
nasal navigation.

In terms of disease reactivation,
there is less tissue disruption en-
do nasally, so the risk of postoperative
inflammation is potentially lower
than with an external approach.
Apart from case 1, no perioperative
steroids were administered be-

Figure 2. Case 2. A, Characteristic saddle nose deformity as well as right orbital inflammation due to active Wegener granulomatosis. B, Coronal computed
tomographic scan demonstrating deviation of the septum to the left side and narrowing of the nasolacrimal duct.

Figure 3. Case 3. A, Collapse of the nasal bridge. B, Coronal computed tomographic scan showing an empty nasal passage with complete destruction of the
ethmoids.

Figure 4. Case 4. A, Axial computed tomographic scan showing a left apical orbital mass causing compressive optic neuropathy. B, No external features of
Wegener granulomatosis. C, Coronal computed tomographic scan demonstrating bilateral medial orbital wall disease causing nasolacrimal duct obstruction.
cause surgery was performed when
the disease was in remission.

While ostium size may be smaller
using an endonasal approach, this
should not increase failure risk, since
WG reactivation would threaten os-
tia of any size. Indeed, external DCR
requires more tissue dissection in the
diseased area, which may increase
the risk of postoperative inflamma-
tion and external scar complica-
tions, such as fistula formation and
wound necrosis. The smaller endo-
anasal ostium may help prevent com-
plications, such as granulomas ex-
tending into the orbit through the
rhinostomy.

Another possible disadvantage of
the endonasal approach is bleeding
from inflamed nasal mucosa obscur-
ing the view, making the surgery
more technically challenging and
causing epistaxis and scarring. In-
flamed nasal mucosa (with hyper-
plasia, erythema, and possible pete-
chiae) suggests active disease and the
surgery should be avoided using
either approach unless the sys-
temic disease is under control, as ad-
vised by the internist based on sys-
temic and laboratory findings. In our
experience, the scarred, necrotic mu-
cosal tissue bleeds less.

While recent radiologic navigating
systems might be considered in cases
where anatomic landmarks are ab-
sent, the standard vitrectomy light
pipe inserted via the superior cana-
lculus into the nasolacrimal sac used
in all 4 cases eliminated any possi-
ble source of confusion and acted as
a target for the surgeon allowing
safe access to the lacrimal sac.

Patients with WG should be
warned about the potential reacti-
vation of their disease and that the
risk of failure is higher because of the
unpredictable nature of their post-
operative inflammation. We recom-
pend preoperative imaging, that an
experienced surgeon perform the
operation, and that systemic dis-
ease is quiescent at the time of sur-
gery; therefore, a multidisciplinary
approach is also advocated. We
would encourage readers to con-
sider the use of EN-DCR in pa-
tients with WG as an equal, and pos-
sibly superior, alternative to the
external approach.

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