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Plastic Surgery Registrar's Conference –April 2013

Vascular Anomalies – High Flow Lesions

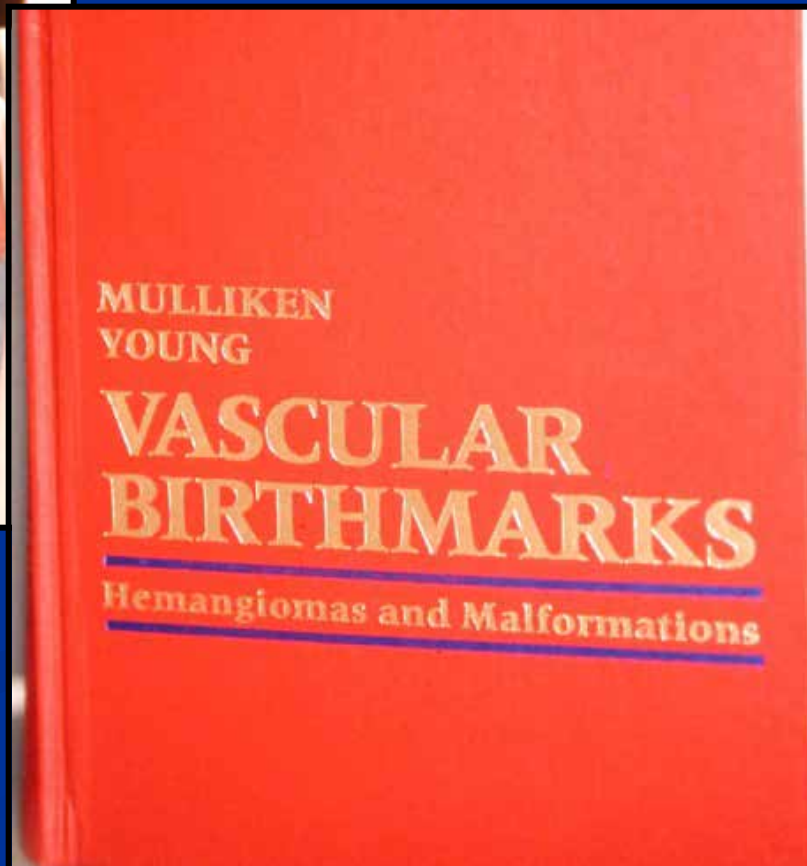
Julian J. Pribaz, M.D.

Nomenclature of vascular anomalies remains problematical in general medical community….

Tumors

v

Malformations


MULLIKEN
YOUNG
VASCULAR
BIRTHMARKS
Hemangiomas and Malformations

Vascular Anomalies: Confusing Terminology

1. Tumors

Biological Name		Incorrect Term
Infantile Hemangioma	→	'Strawberry Hemangioma' 'Capillary Hemangioma' 'Cavernous Hemangioma'
Congenital Hemangioma	→	Infantile Hemangioma
Hemangioendothelioma	→	'Capillary Hemangioma'
Pyogenic Granuloma	→	'Hemangioma"

Vascular Anomalies: Confusing Terminology

2. Malformations

Biological Name	Incorrect Term
Capillary Malformation	“Port-wine stain” “Capillary hemangioma”
Lymphatic Malformation	“Cystic hygroma” “Lymphangioma”
Venous Malformation	“Cavernous hemangioma”
Arteriovenous Malformation	“Arteriovenous hemangioma”

Vascular Anomalies- Biological Classification:

Clinical & Endothelial Characteristics

Hemangiomas v Malformations

- Proliferating phase
- Involuting phase

- Capillary
- Venous
- Arterial
- Lymphatic
- AV Fistulae(AVM)

Mulliken JB, Glowacki J. PRS 1982

International Society for Study of Vascular Anomalies (ISSVA) 1996

Tumors

Hemangioma
Hemangioendotheliomas
Angiosarcoma
Miscellaneous

Malformations

Slow-Flow
Capillary
Lymphatic
Venous

Fast-Flow
Arterial
Combined

Vascular Anomalies 2: Malformations –slow flow

Capillary Malformation (CM)

Lymphatic Malformation (LM)

Microcystic & Macrocystic Lymphedema

Venous Malformation (VM)

Combined – CVM, CLM, CLVM, LVM

Vascular Anomalies 2: Malformations –Fast flow

Arterial Malformation (AM)

Aneurysm, Atresia, Ectasia, Stenosis

Arteriovenous Malformation (AVM)

Combined

CAVM , CLAVM

Epidemiology of Vascular Anomalies

General Population

Hemangioma (~4%)

Malformations (~0.5%)

Referral to Vascular Anomalies Center

Boston Childrens Hospital (n=5621)

Tumors (35.2% (n=1976)

Infantile Hemangioma	85.9%
Hemangioendotheliomas	7.8%
Congenital Hemangioma	5.4%
Pyogenic Granuloma	0.9%

Malformations (64.8% (n=3645)

Venous	36.8%
Lymphatic	28.3%
Arteriovenous	14.3%
Capillary	11.0%
Combined	9.6%

Greene, et al. J Ped Surg 2011

AVMs of Head and Neck

- Poorly understood
- Difficult to treat

Arteriovenous Malformation of The Head and Neck: Natural History and Management

Kohout, M.P., Hansen, M., Pribaz, J.J, Mulliken, J.B.
P.R.S. 102: 643-654 (1998)

Results overview

- 81 Patients (33M : 48F)
- M:F = 1 : 1.5
- Age at presentation: 3 months – 66 years
- Mean follow-up = 4.6 years

Clinical Presentation & Course

Depends On:

- Position, size, number, length of abnormal AV connections
- Hemodynamics
  - high flow
  - low flow

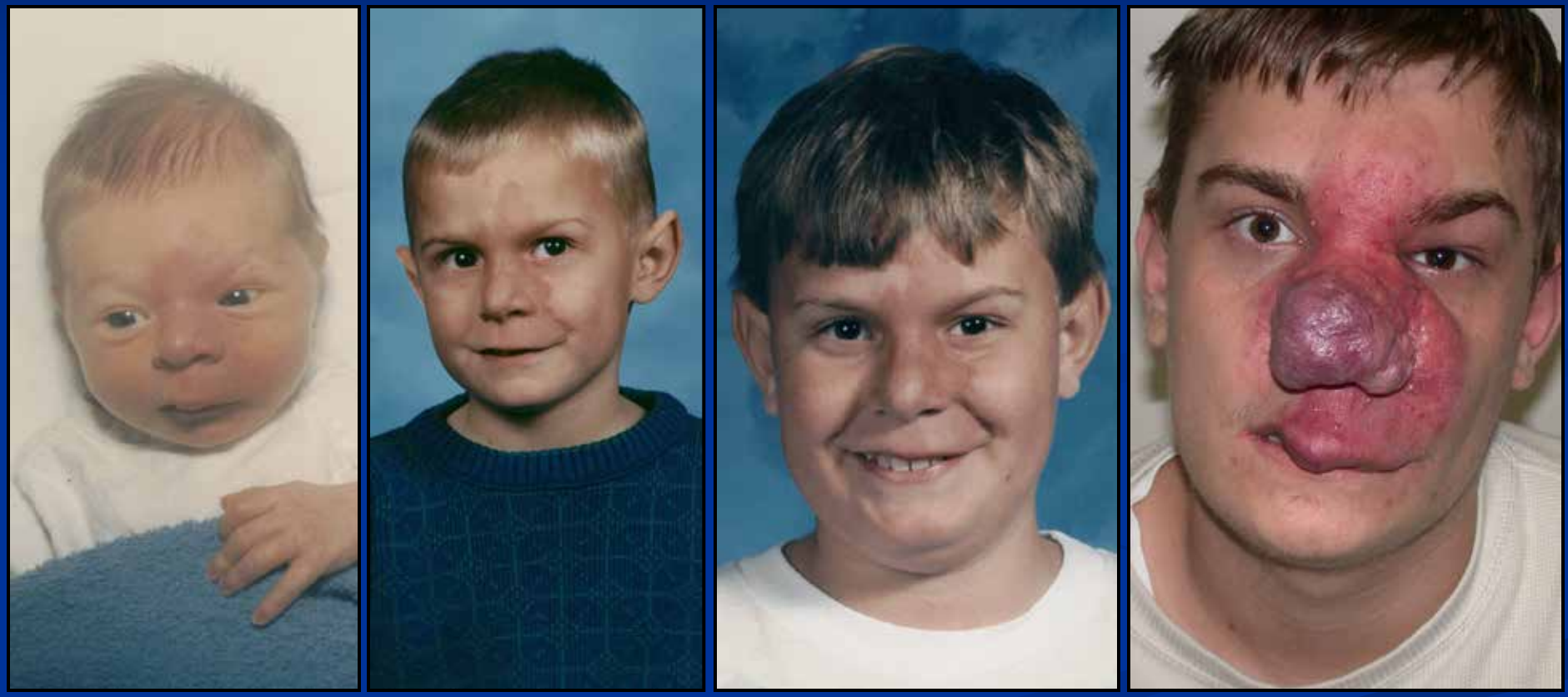
Progression of AVM

Age 16

Age 22

Age 34

Progression of AVM



Newborn Age 5 Age 10 Age 16

AVM – Head and Neck
Age of Onset

Birth	-	59% (M>F)**
Childhood	-	10% (F>M)
Adolescence	-	10% (F>M)
Adult	-	21% (F>M)

AVM – Head and Neck Distribution

Midface	-	69%**
Upper 1/3	-	14%
Lower 1/3	-	17%

Schobinger Clinical Staging at Presentation

STAGE I (Quiescence)	**27%**
STAGE II (Expansion)	**36%**
STAGE III (Destruction)	**36%**
STAGE IV (Decompensation)	**1%**

AVM – Causes of Ulceration

1. **Arterial ischemia**
   - prox "steal"
2. **Venous hypertension**
   - (like VVs)
   - fibrinogen leak
   - precapillary fibrin cuffing

   ⇩ perfusion of O_2 & nutrients

   (Barnard & Browse, 1982)
3. **Trauma**
4. **2º ill-advised XRT, sclerotherapy, prox lig**

Ischemic ulcers –very painful

Clinical Presentation

Asymptomatic

- Mass
- Deformity
- Bruit

Symptomatic

- Pain
- Bleeding
- Ulceration

stable → progressive

Precipitating Factors - AVM

Trauma

- 2 falls
- 14 iatrogenic insults (bx, lig, exc)

Puberty
F>M

- 8 started (2M, 6F)
- 6/31 expanded (2M, 4F)

Pregnancy

- 4 started
- 11 expanded – esp late onset AVM
- aggressive
- partial resolution post partum.

Timing of surgical treatment depends on severity of symptoms.

Treatment

Asymptomatic → observe

Symptomatic/ & Deformity → treat

(a) small AVM → wide local excision

(b) large AVM → **selective** embolization
resection (2-14d later)
& reconstruction

Patients are usually young & have very high expectations

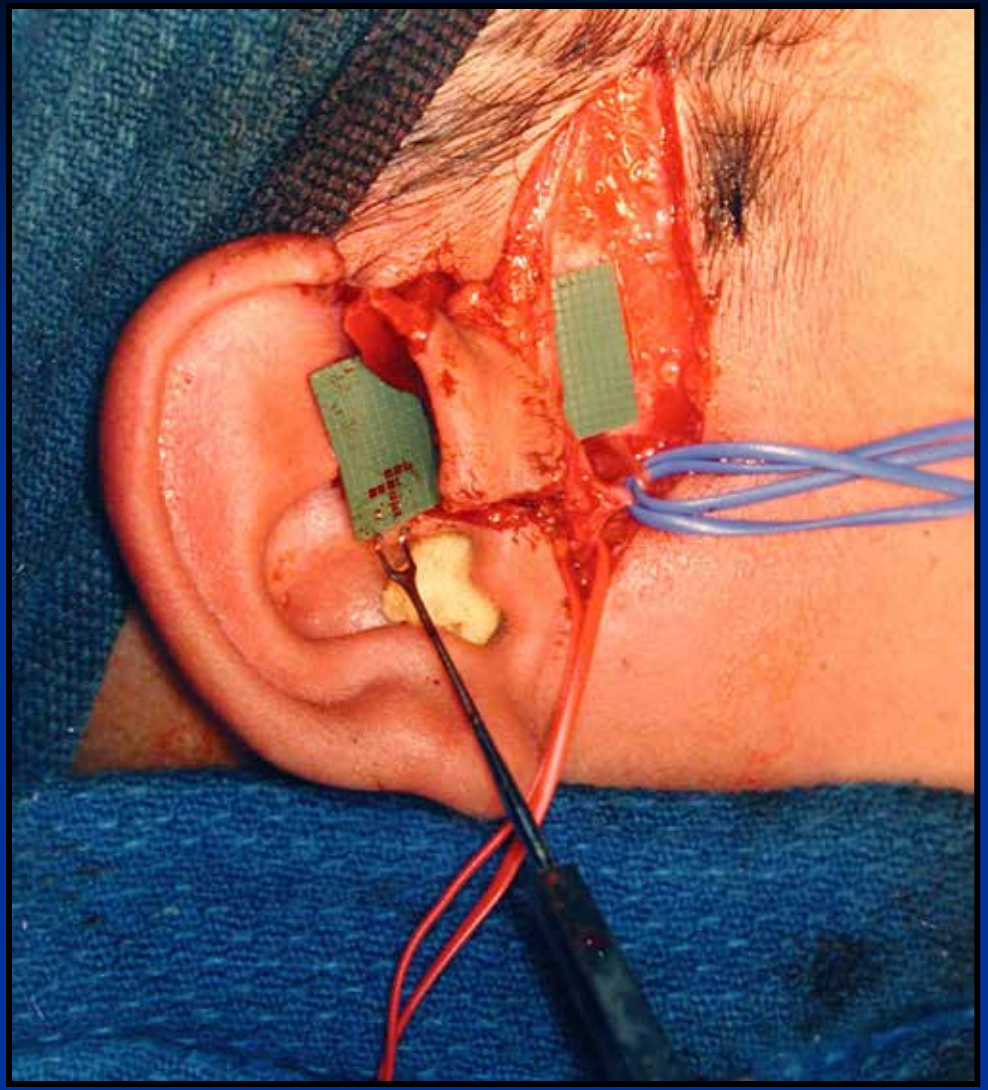
SMALL AVM

Reconstruction with laminated free flap

5M – AVM (L) ALAR

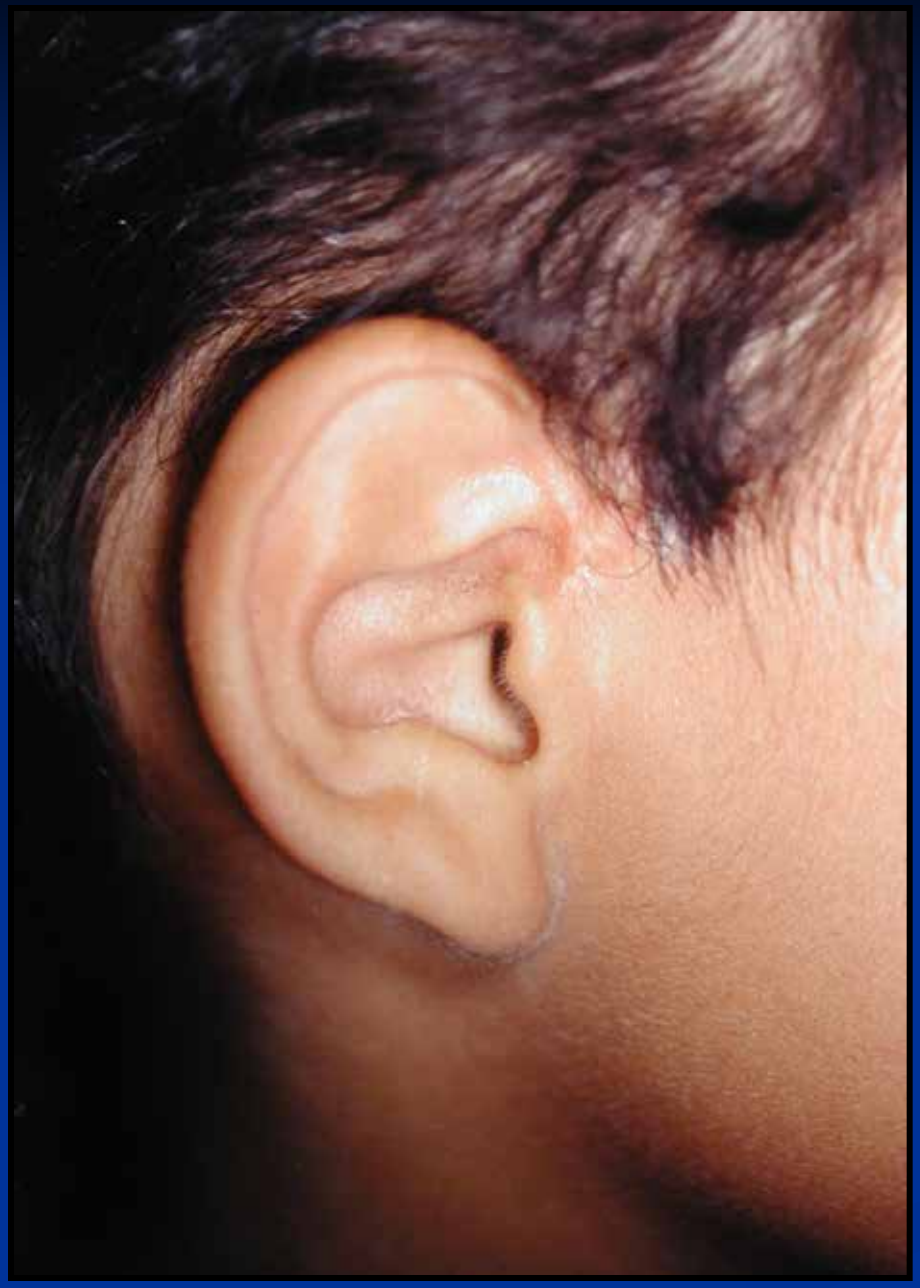
Cadaver dissection of nose & ear

- ascending helix of ear g distal heminasal reconstruction

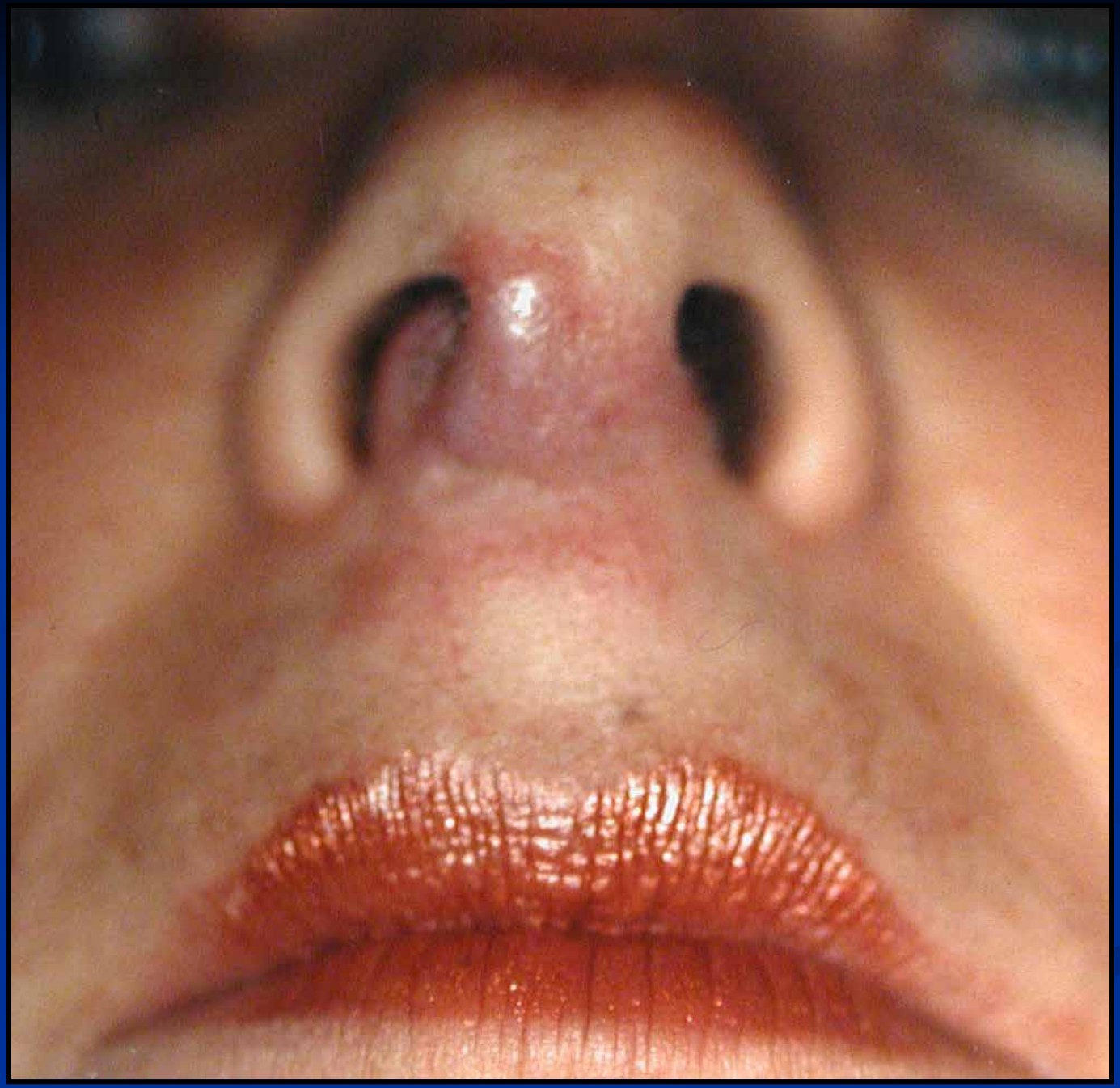


Ascending helical flap dissection
based on sup. temporal vessels

Flap transfer
Anastamsis to facial A&V



6 months post op



AVM of nose & upper lip

41F AVM of nose & upper lip

AVM resection

Ascending helical flap from (R) ear to columella

Bone wax model of defect

6 months post op

Prior Treatment of AVMs 50%

Steroids

Irradiation

Laser

Embolization

Subtotal resection

Investigation of AVM

- Doppler ultrasound
- MRI (soft tissue)
- CT (bone)
- Angiography
- MRA

Treatment of AVMs of Head and Neck

TEAM APPROACH

Embolization	Resection	Reconstruction
Reduces bleeding, not extent of resection	?Cure/Control	Restore appearance and function

Problems with AVM Treatment in Head and Neck

Embolization/Excision – I

- ? Endothelial injury from embolization
- Blood loss (esp. bone)
- ? Complete excision

Excision Margins

- Bleeding pattern
- Doppler
- ? Histology

Problems with AVM Treatment in Head and Neck

Embolization/Excision – II

- Massive defect
- Complex 3D defects
- Young patients
- Complications from previous Mx (prox lig. XRT)

AVM

? Proximal ligation of AVM – ineffective

prox. ligation

È

distal ischemia

↓

↑ **collaterals**

↓

↑ **flow through fistula**

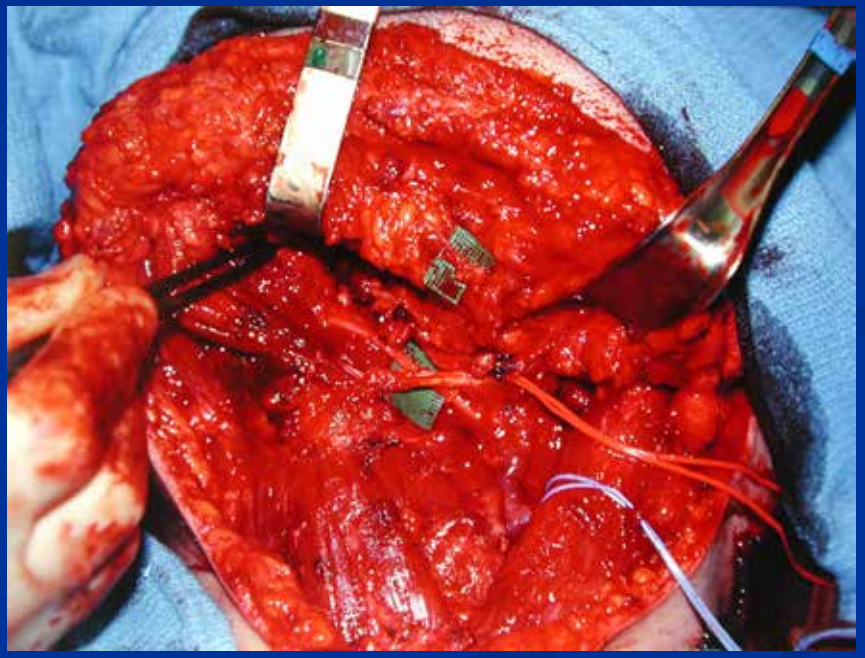
AVM

Localized

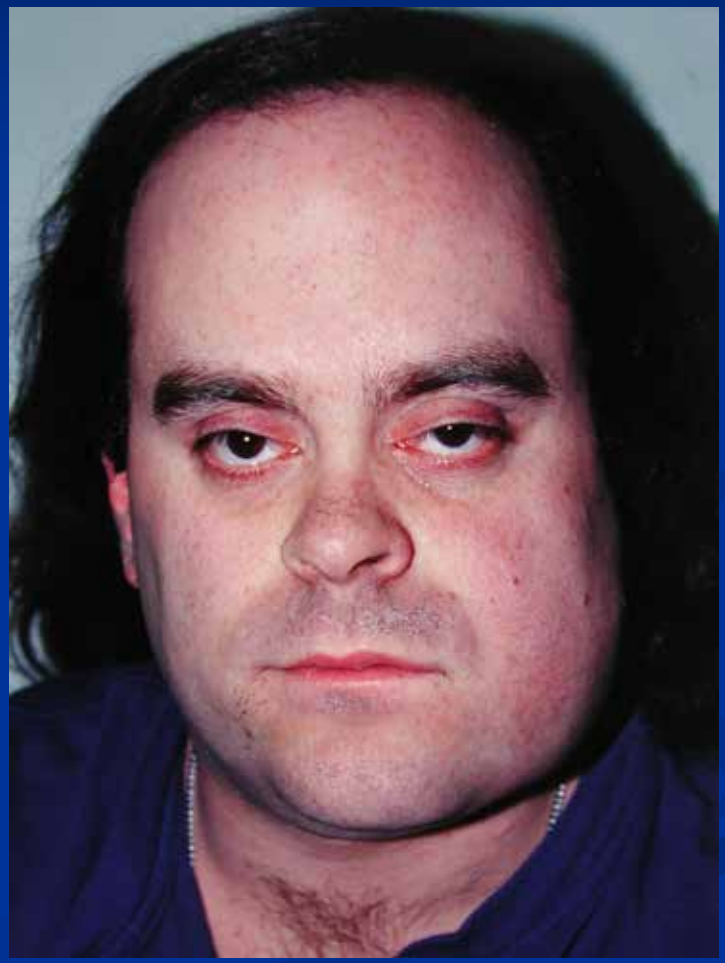
Localized low flow AVM

(reversed)

Wide excision AVM

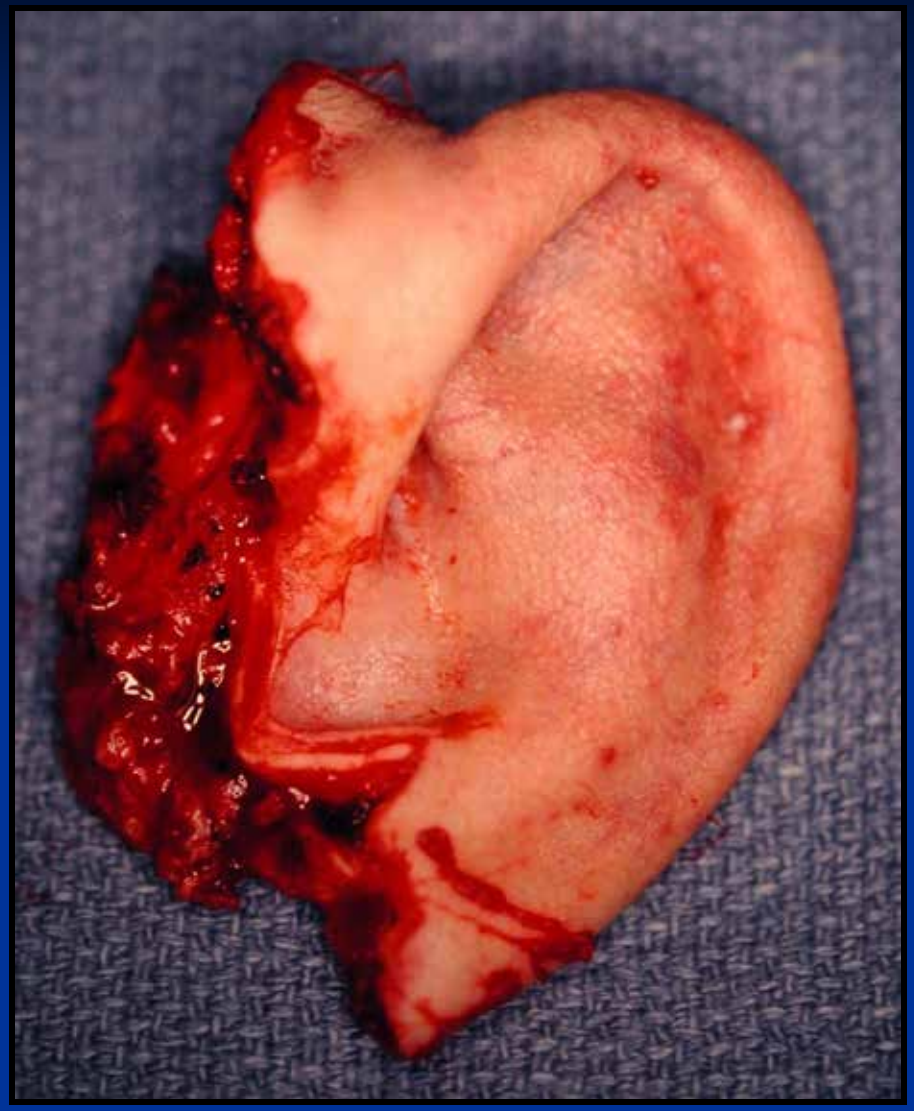


Post operative



Localized high flow AVM

15 yr old male with AVM of ear

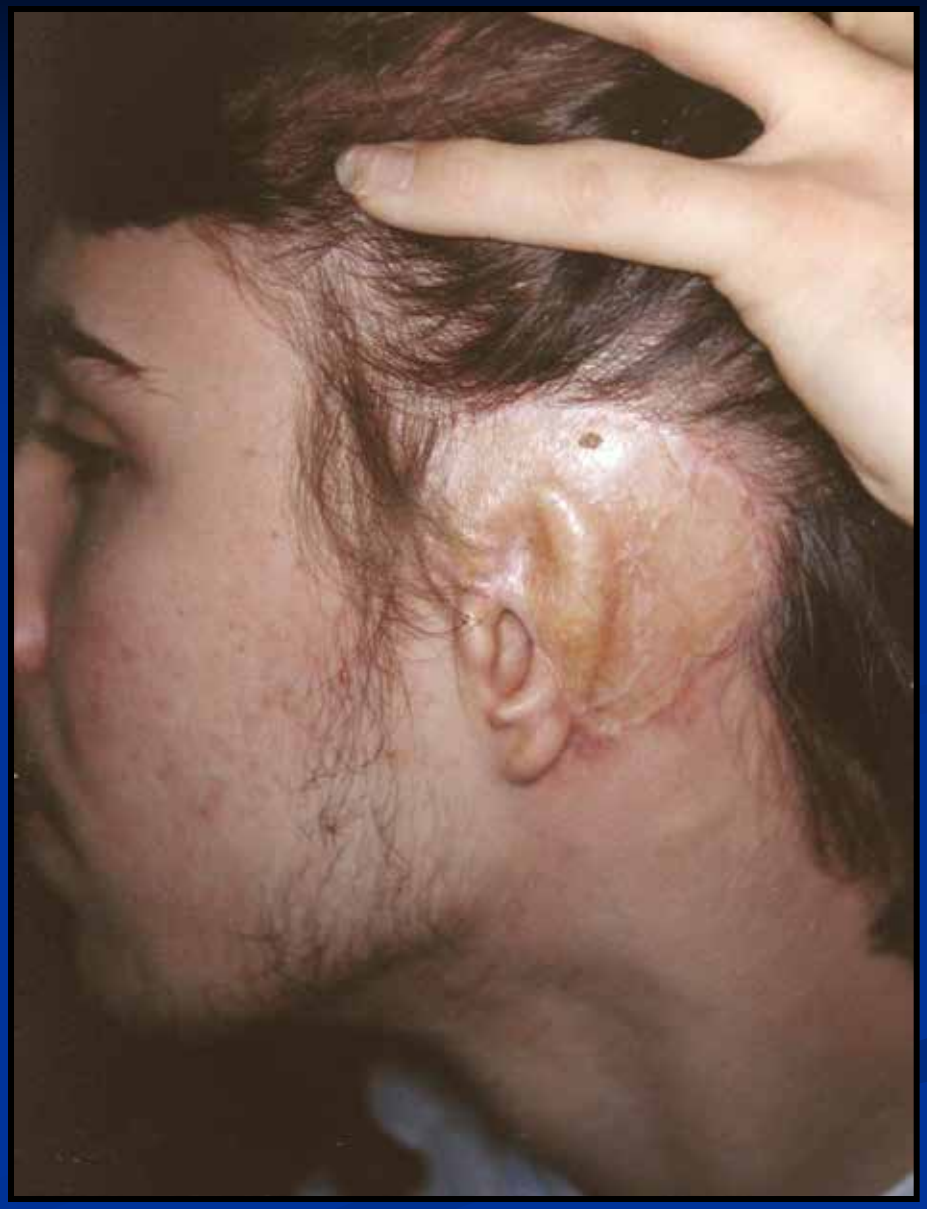


Resected AVM of ear

Avascular auricular cartilage free of AVM

Ear cartilage reattached and TPF flap applied

FTSG over TPF flap



9 months post op

Extensive low flow AVM

Post embolization

Early post operative after radical debulking of low flow AVM

preoperative

After 2 debulking procedures of low flow AVM

Radical excisions of high flow AVMs

create complex defects

Reconstruction of Complex Defects with Tailored Free Flaps

Techniques:

- **Modeling**
  - **alginate**
- **Flap manipulation**
  - **tissue expander**
  - **compound flap on 1 pedicle**
  - **folded flaps**
- **Staged reconstruction**
  - **2º local pedicle flaps**
  - **prelaminated flaps**

Problems with AVM Treatment in Head and Neck

Free Flap Reconstruction - I

- **Recipient vessel injury**

  (prox. lig & embolization injury)

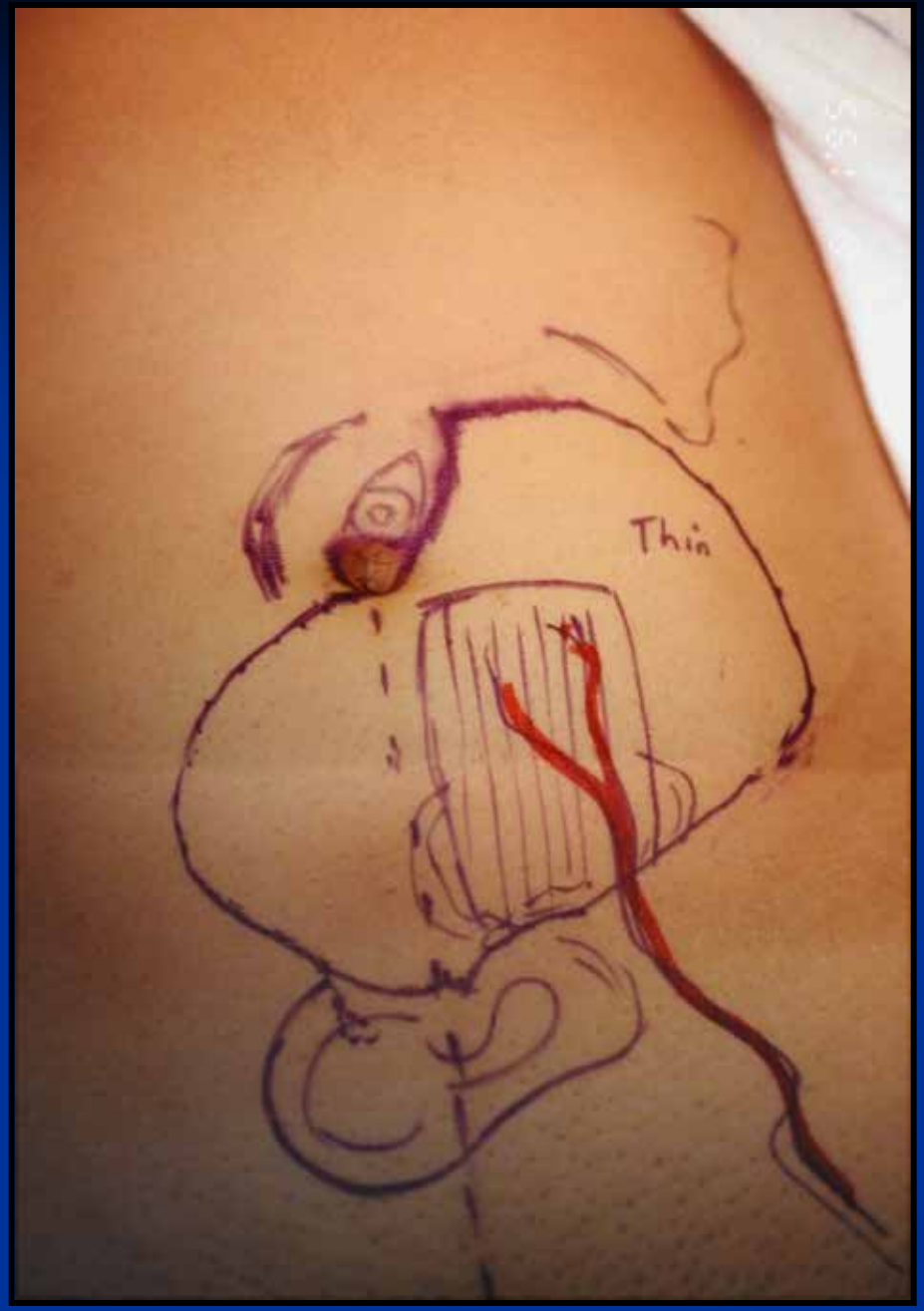
- **Complex 3D flap reconstruction**
  - **laminated**
  - **prelaminated**
  - **preexpanded**
  - **folded**
  - **staged**

Problems with AVM Treatment in Head and Neck

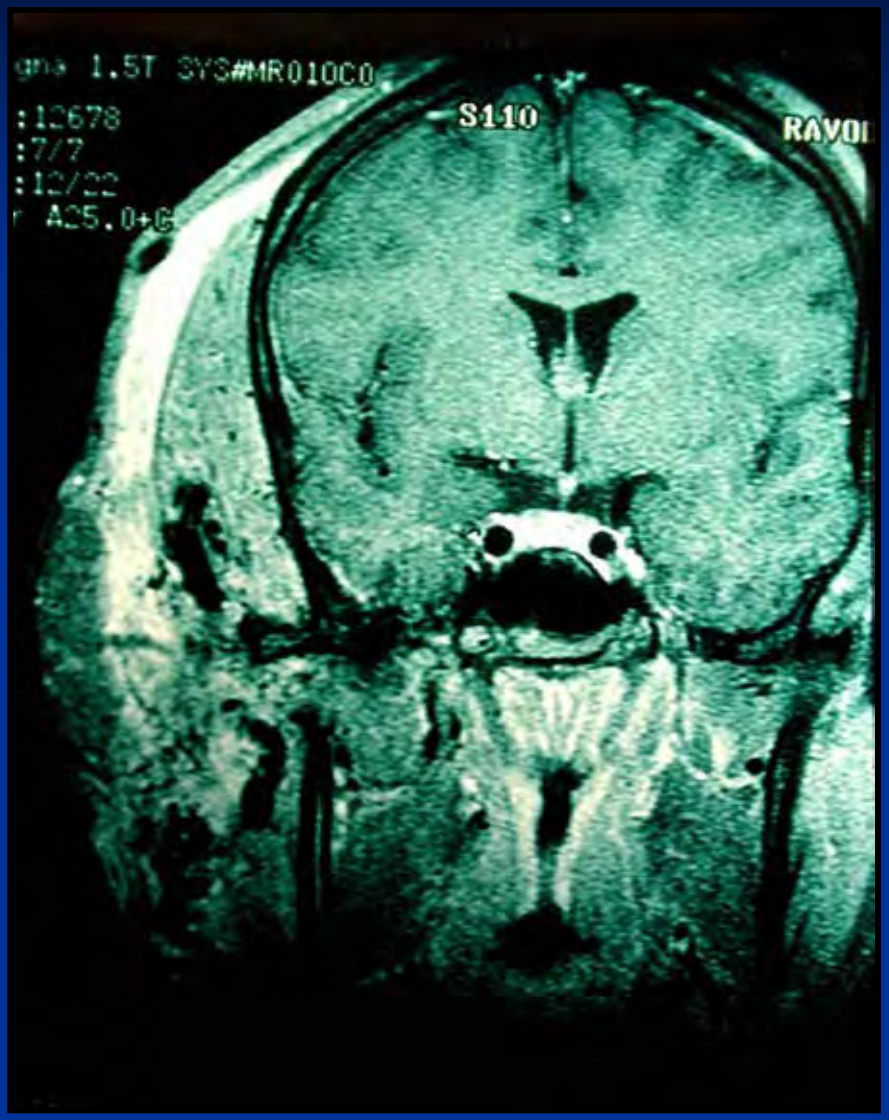
Free Flap Reconstruction - II

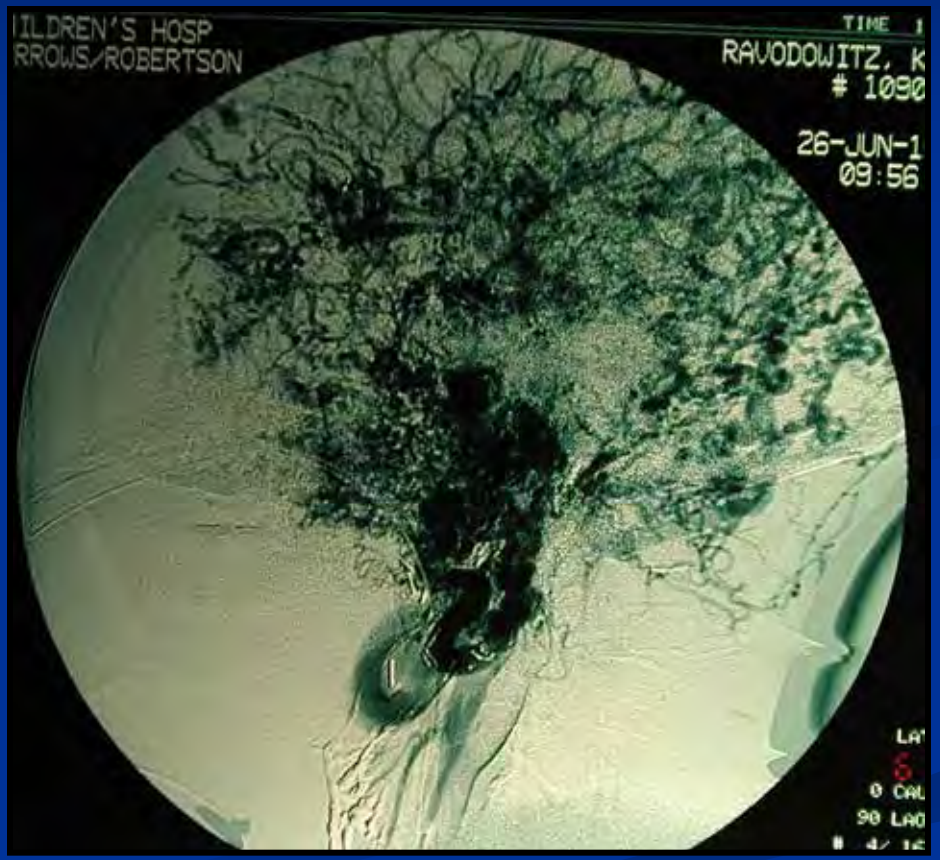
- **Functional restoration**
  - **Muscle**
  - **Bone/teeth**
- **Aesthetic refinements**
  - **Multiple revisions**
  - **Local flaps**

19M – extensive ulcerated AVM face

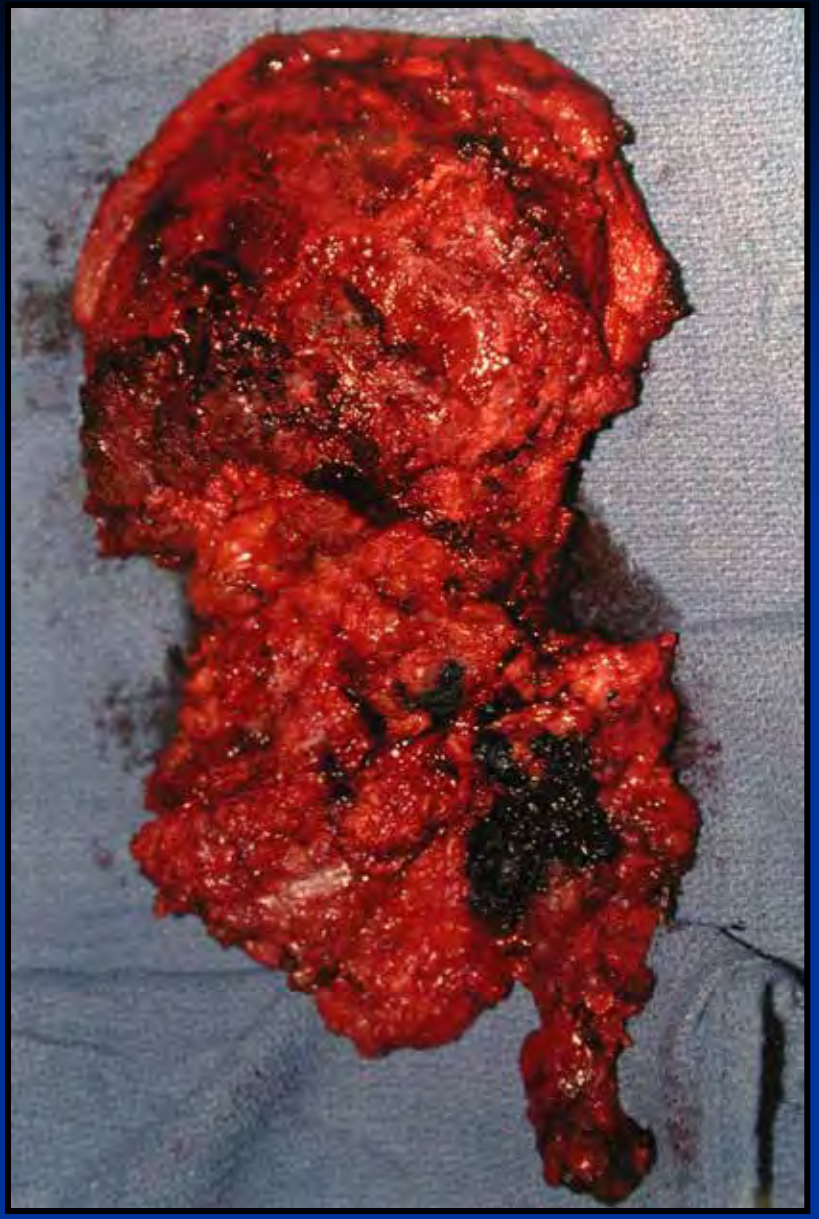


Reconstruction with free TRAM flap



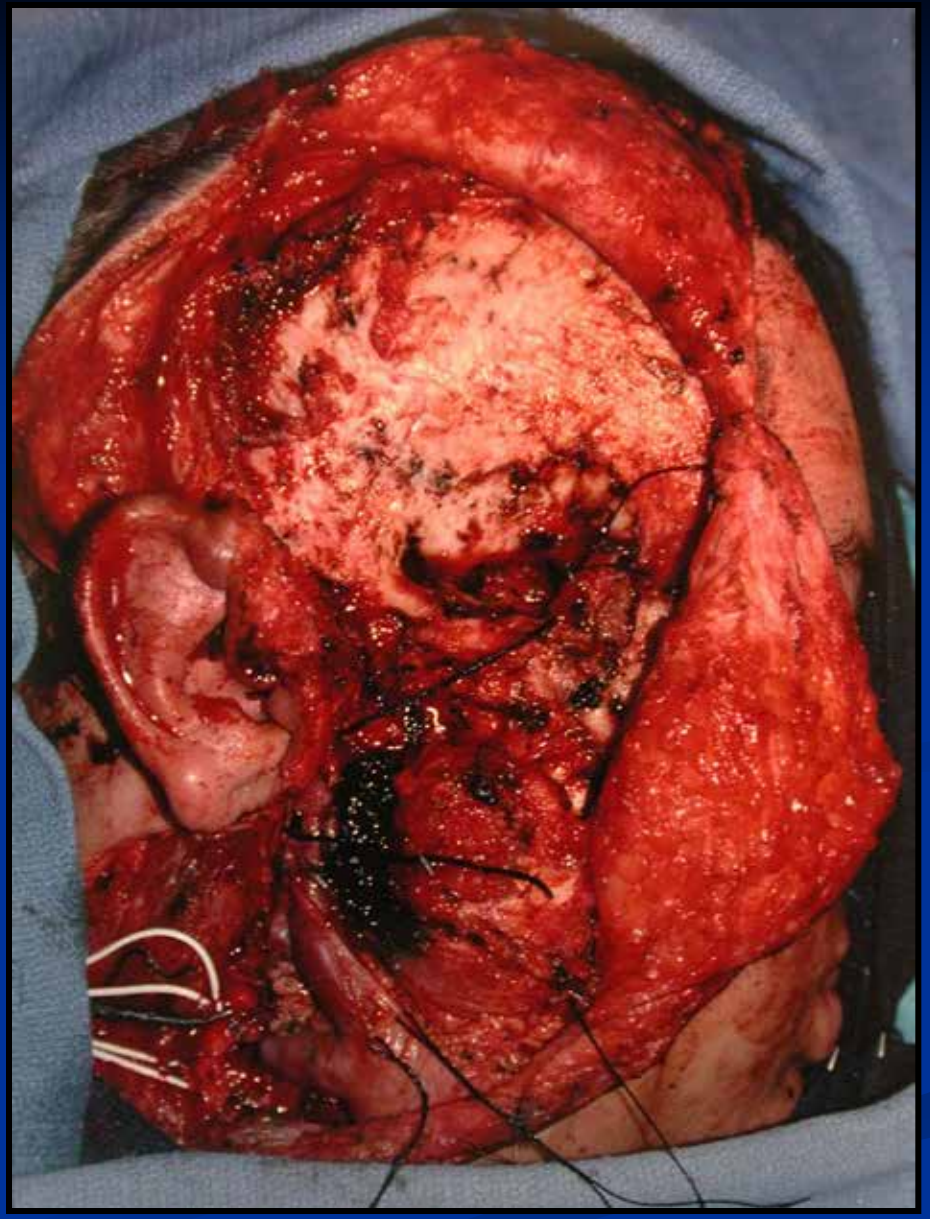


Recurrent/Residual AVM in temporal fossa, masseter & parotid



Excised specimen

-Temporalis M
-Parotid & facial N.
-Masseter M.

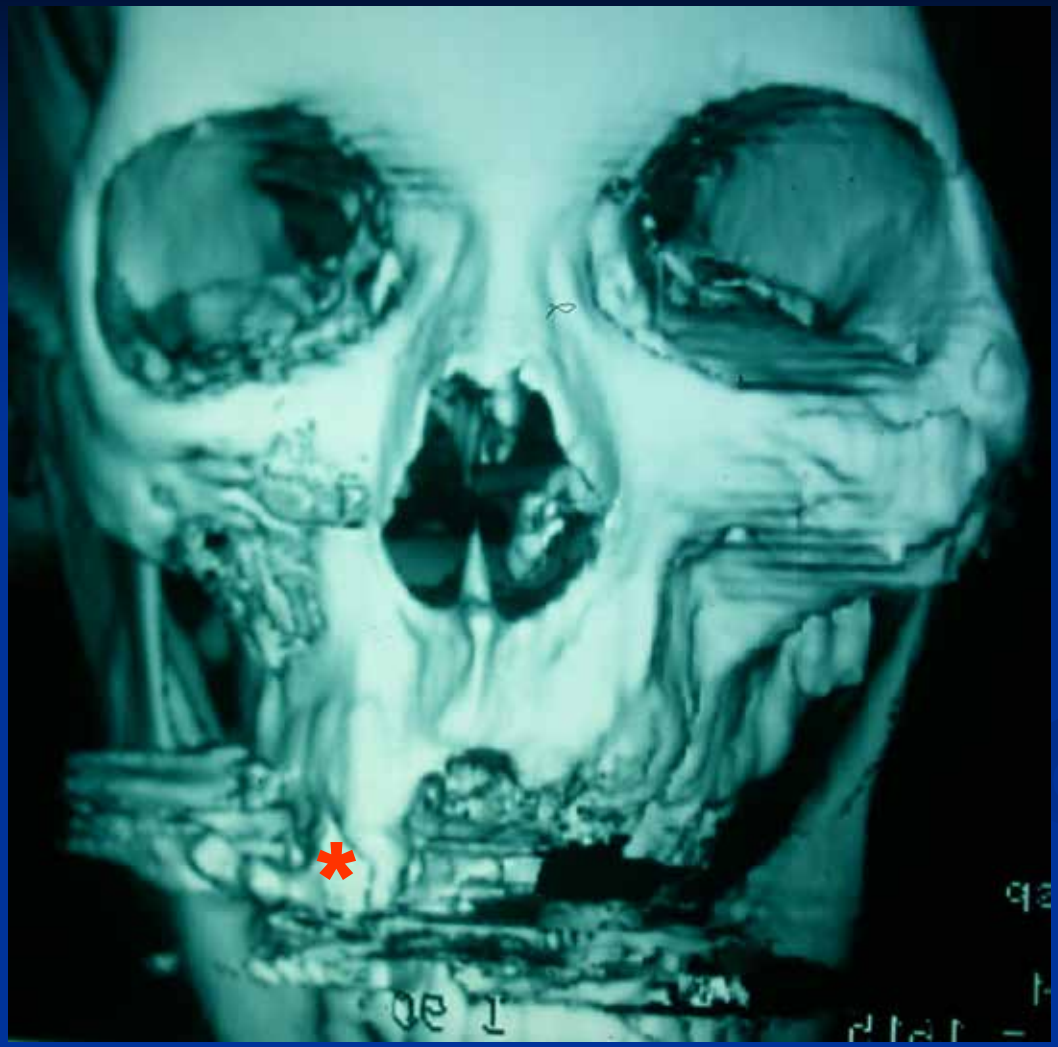


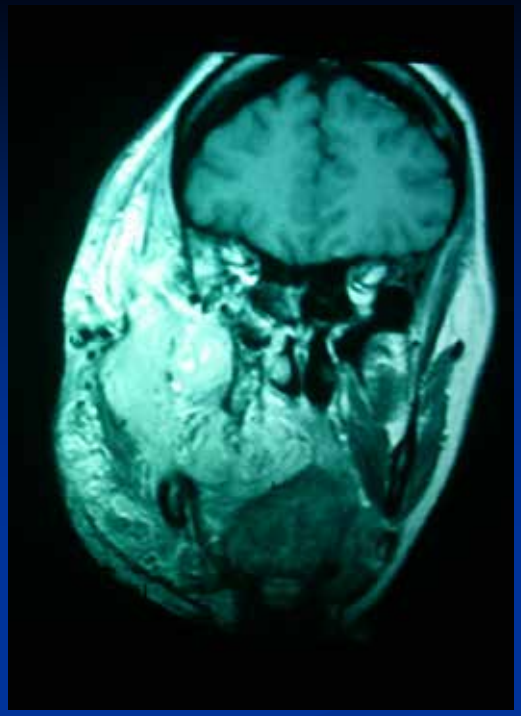
Facial nerve graft

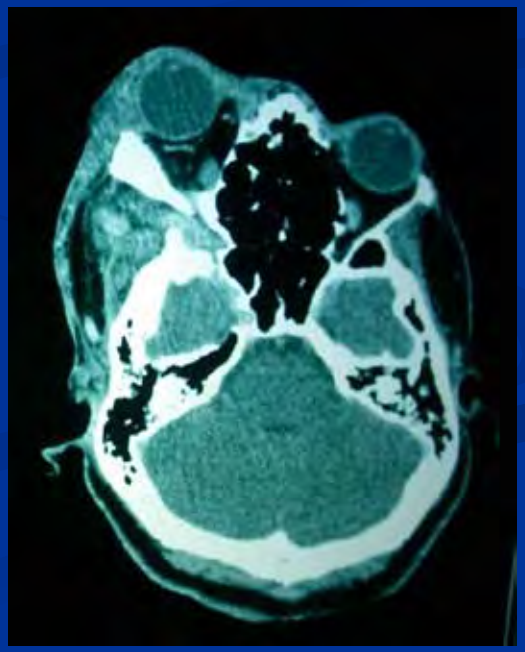
4 months post op

34 F AVM R orbit, maxilla, cheek, facial m, lips

Overgrowth R maxilla

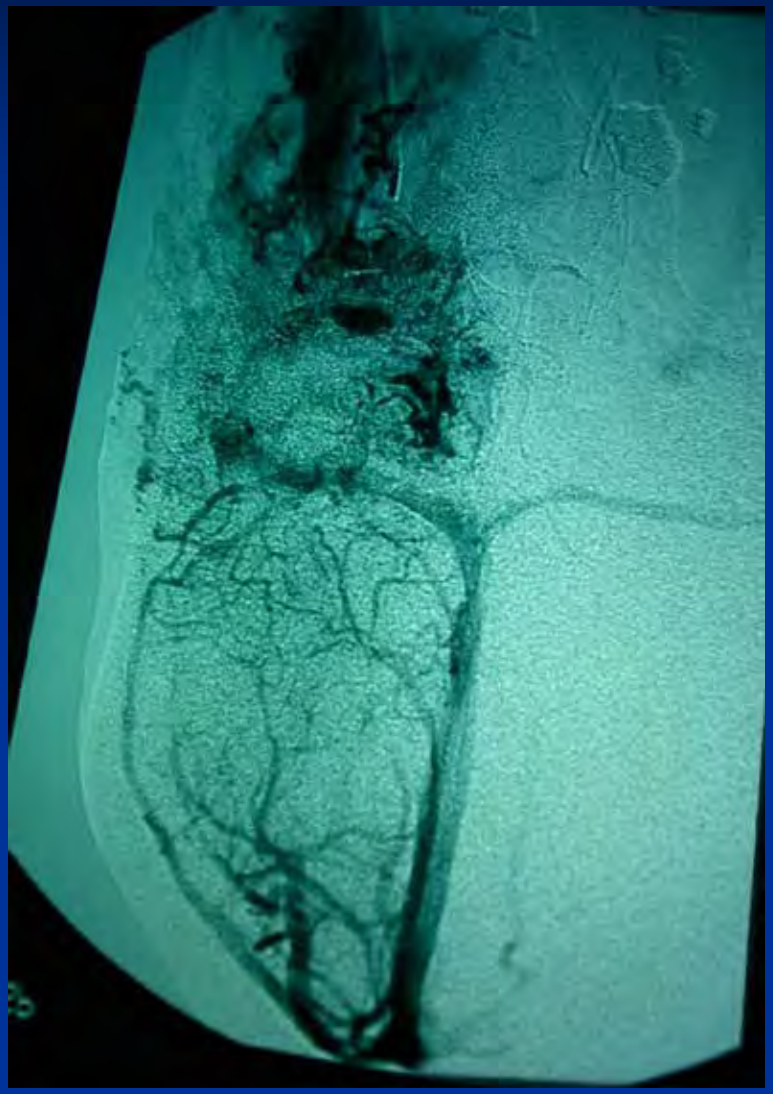


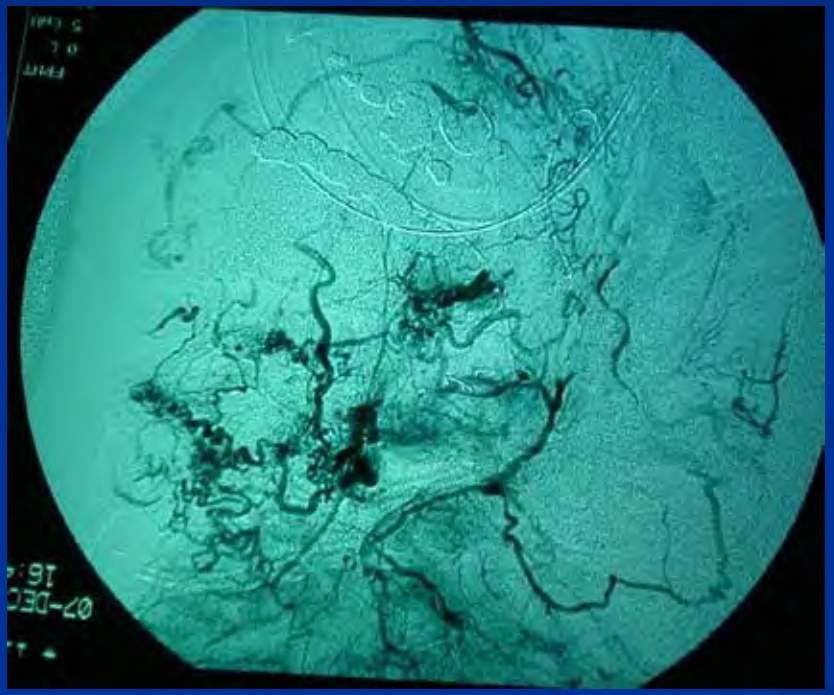




Overgrowth R maxilla

embolization





AVM resected

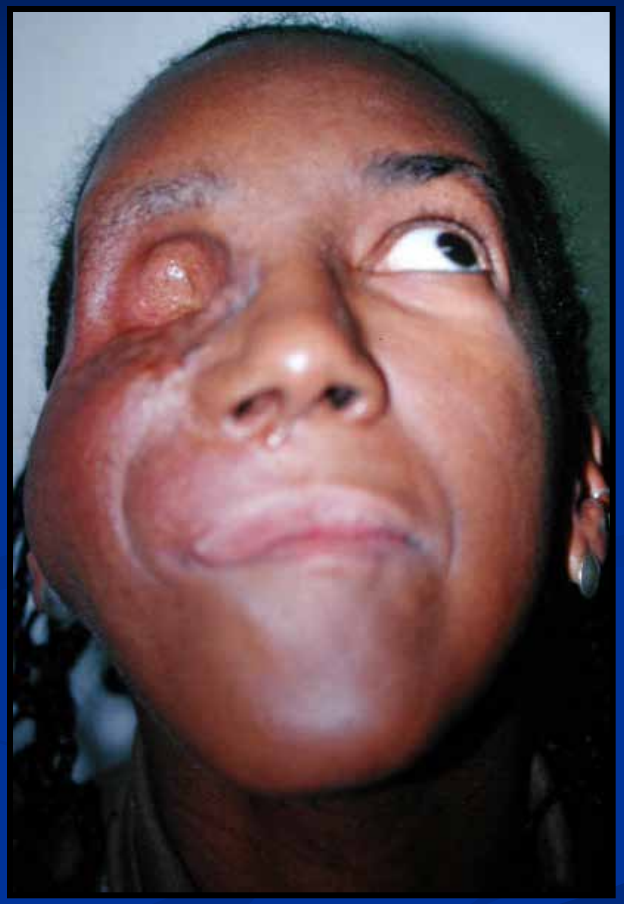
Hemi-Lefort 1 osteotomy(WS)

Planned functional rectus abd m/c flap

R eye left per patient request

Early post-op

6 m later - exenteration of R orbit & flap revised



Pre op

2 year post op

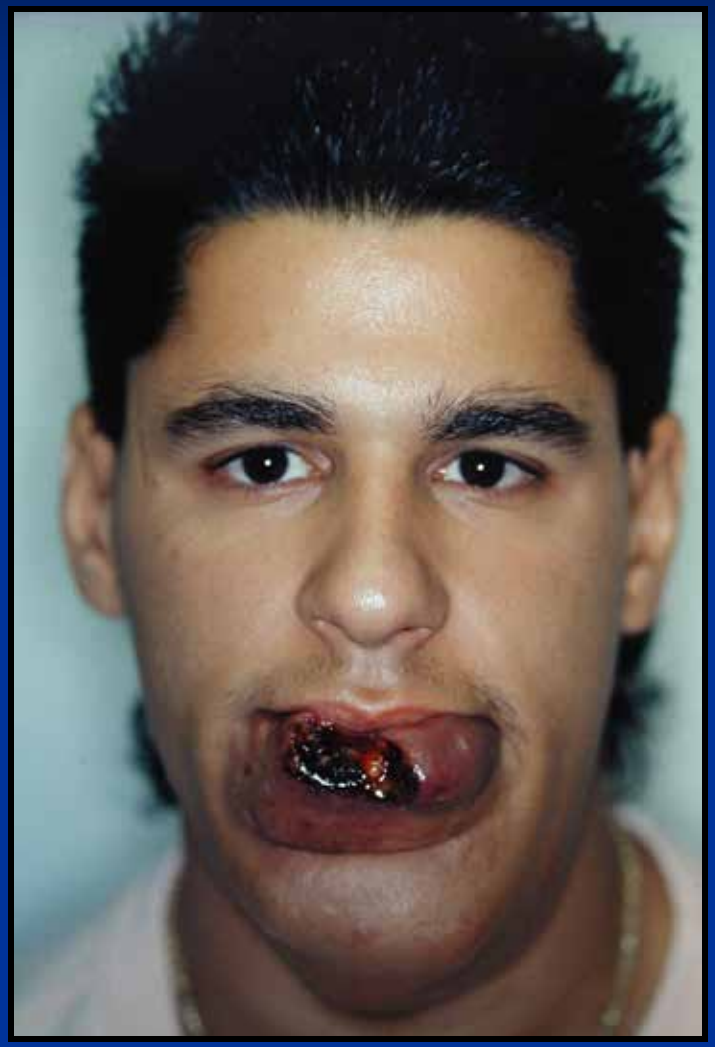
Microsurgical Reconstruction of Head & Neck

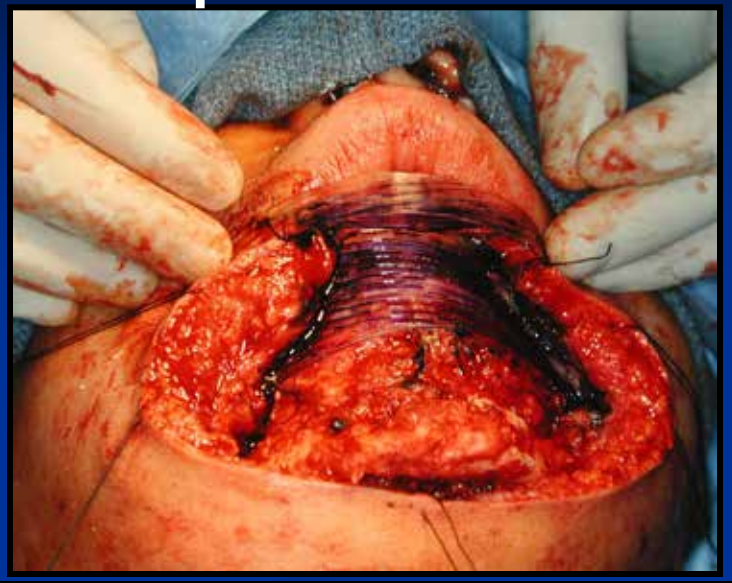
Multifaceted & Laminated Flaps

Folded Flap

Folded Free Flap

Ulcerated AVM of lower lip & chin





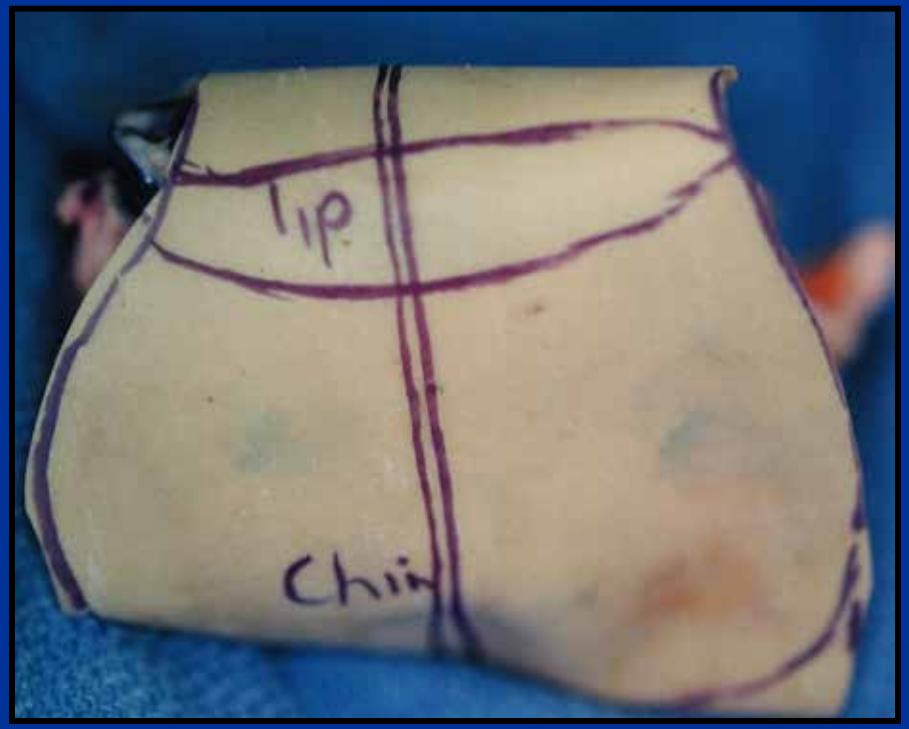
Intra-operative alginate model of the defect

3-D MODEL

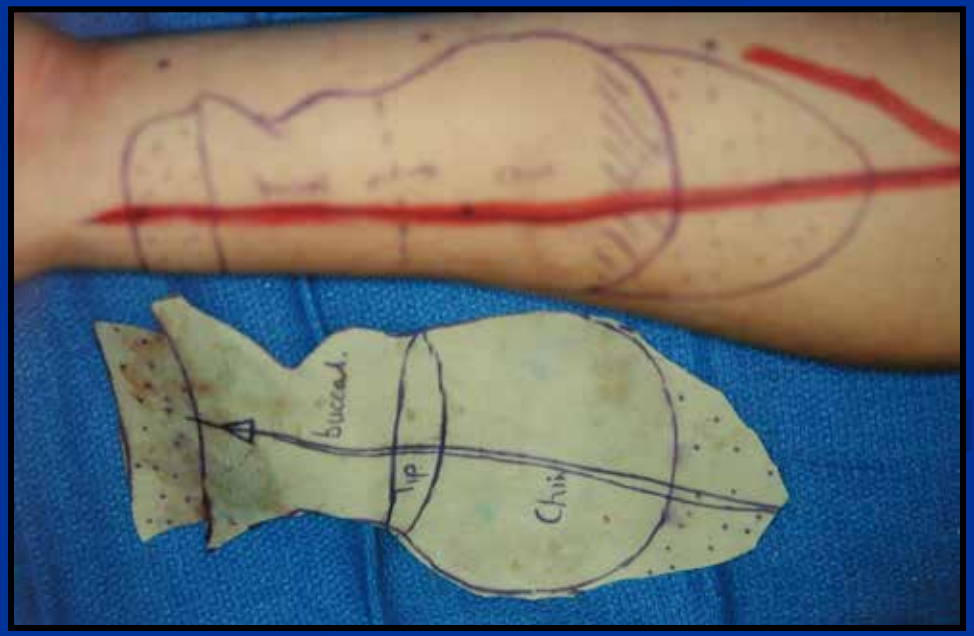
2-D PATTERN

VOLUME

COVER/LINING (S)

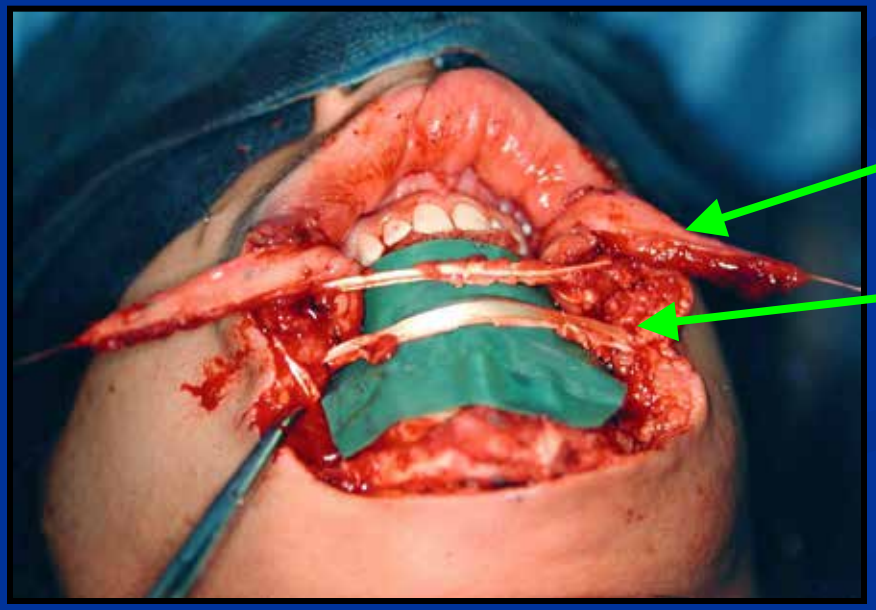


Tailored radial forearm flap



Pribaz,Morris, Mulliken PRS 1994

Bilateral FAMM flaps for vermillion reconstruction

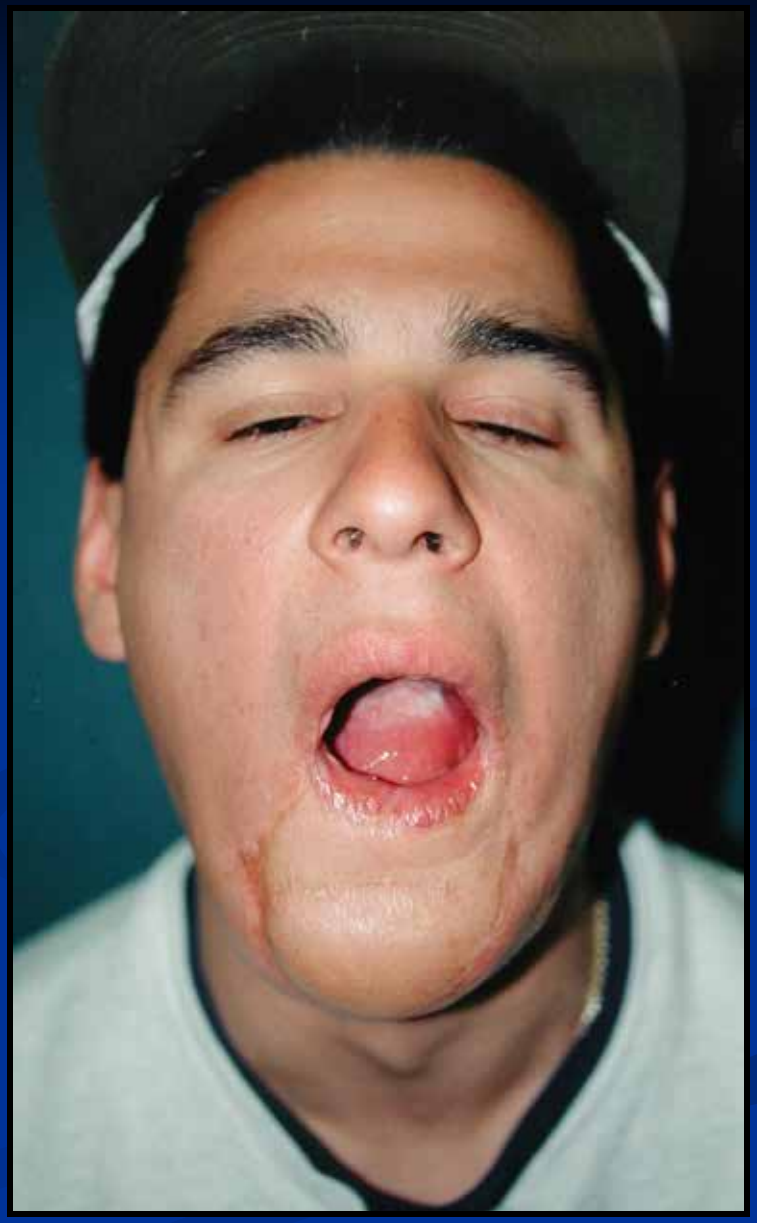


FAMM flap

Oral sling with palmaris longus tendon graft

(intact modiolus)

Post – folded radial forearm flap and bilateral FAMM flap



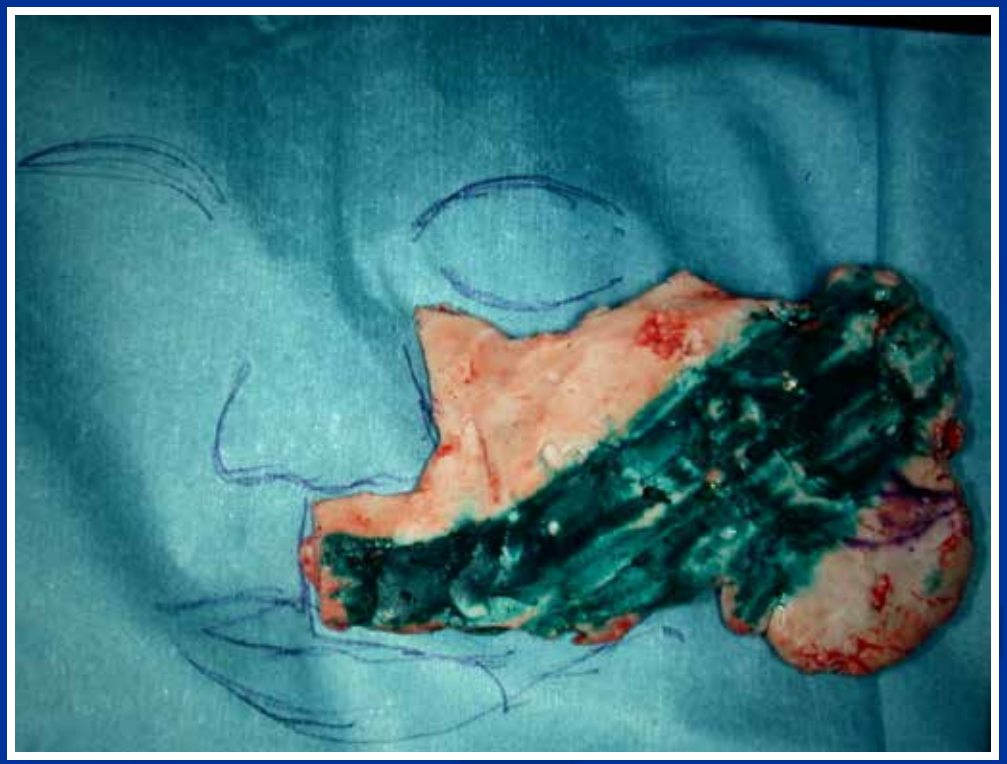
4 years post op

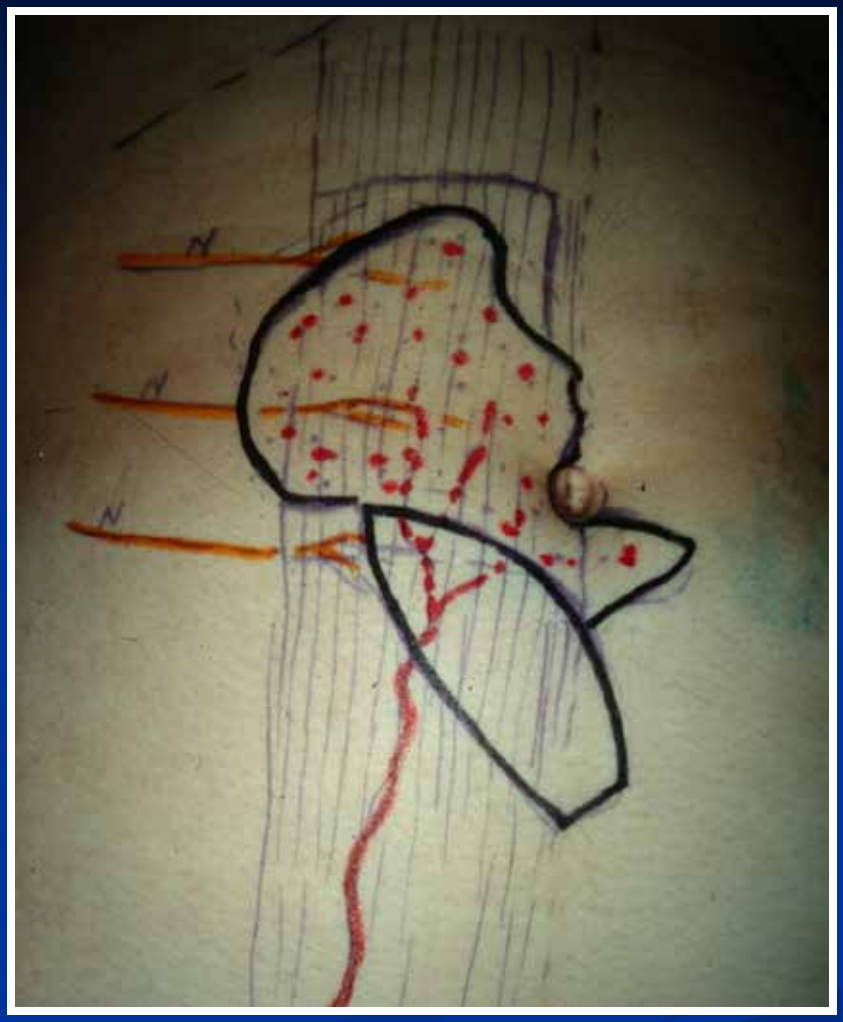
20 F extensive AVM of L maxilla & cheek

Resection AVM :

buccal mucosa, facial m, partial left max

Alginate model of defect





3D model used to design tailored flap from rectus abd. flap with planned seg nerve harvest

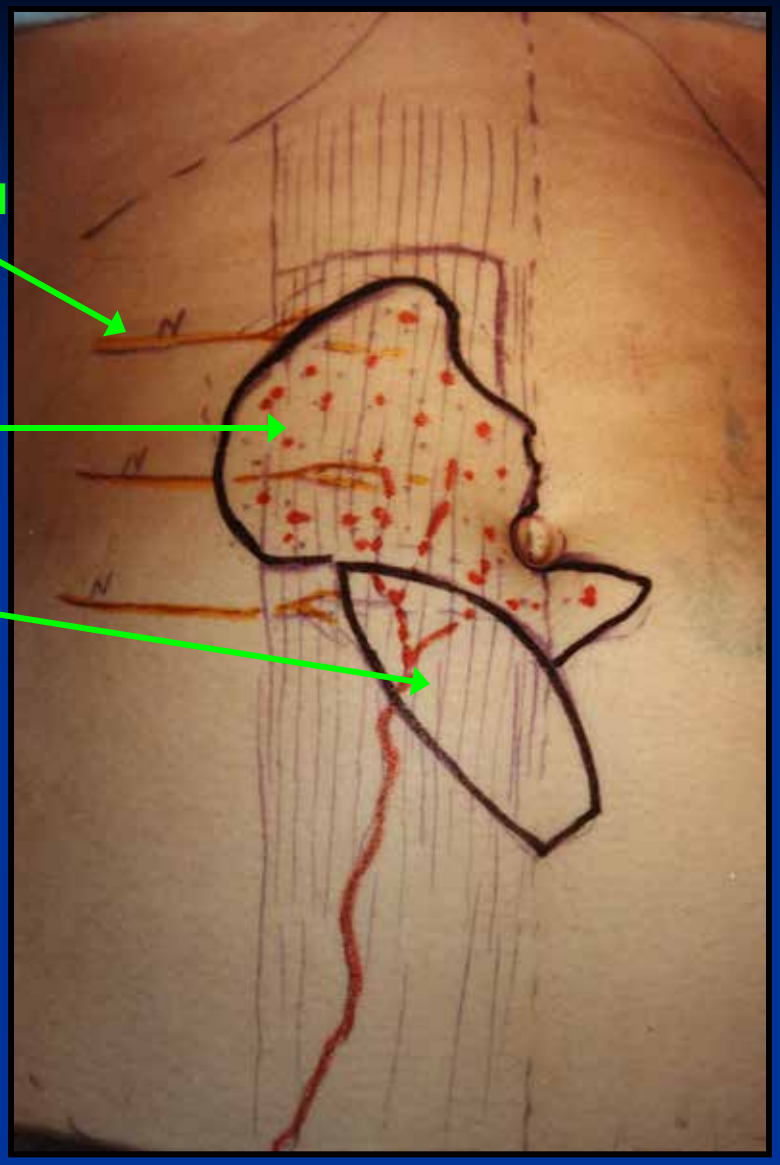
Alginate model showing direction of needed muscle pull for smile

Rectus abdo. M/C flap planned

Intercostal nerves

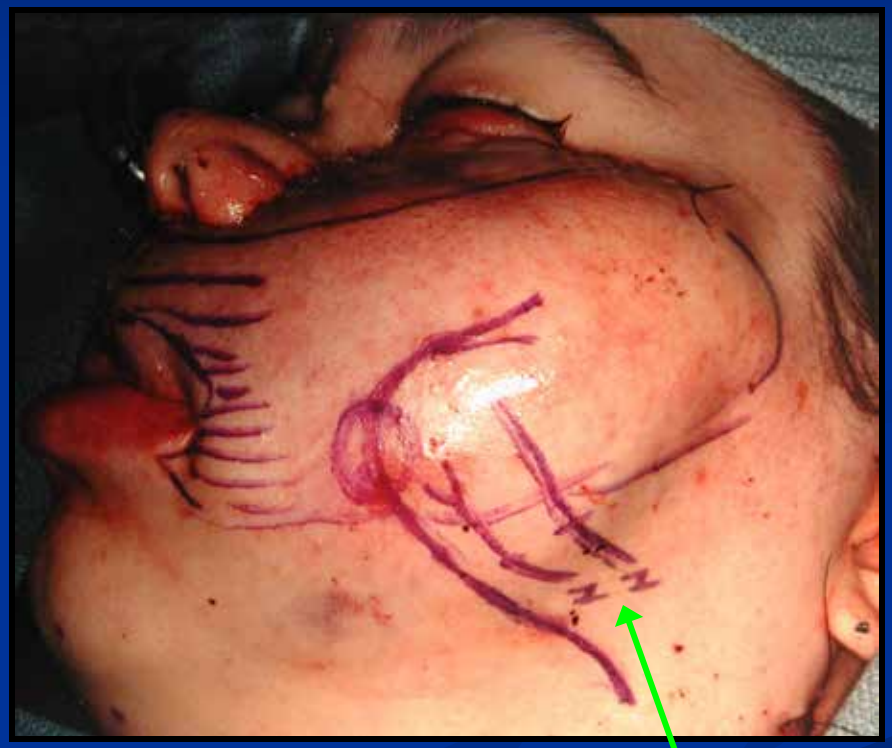
De-epith

Skin island

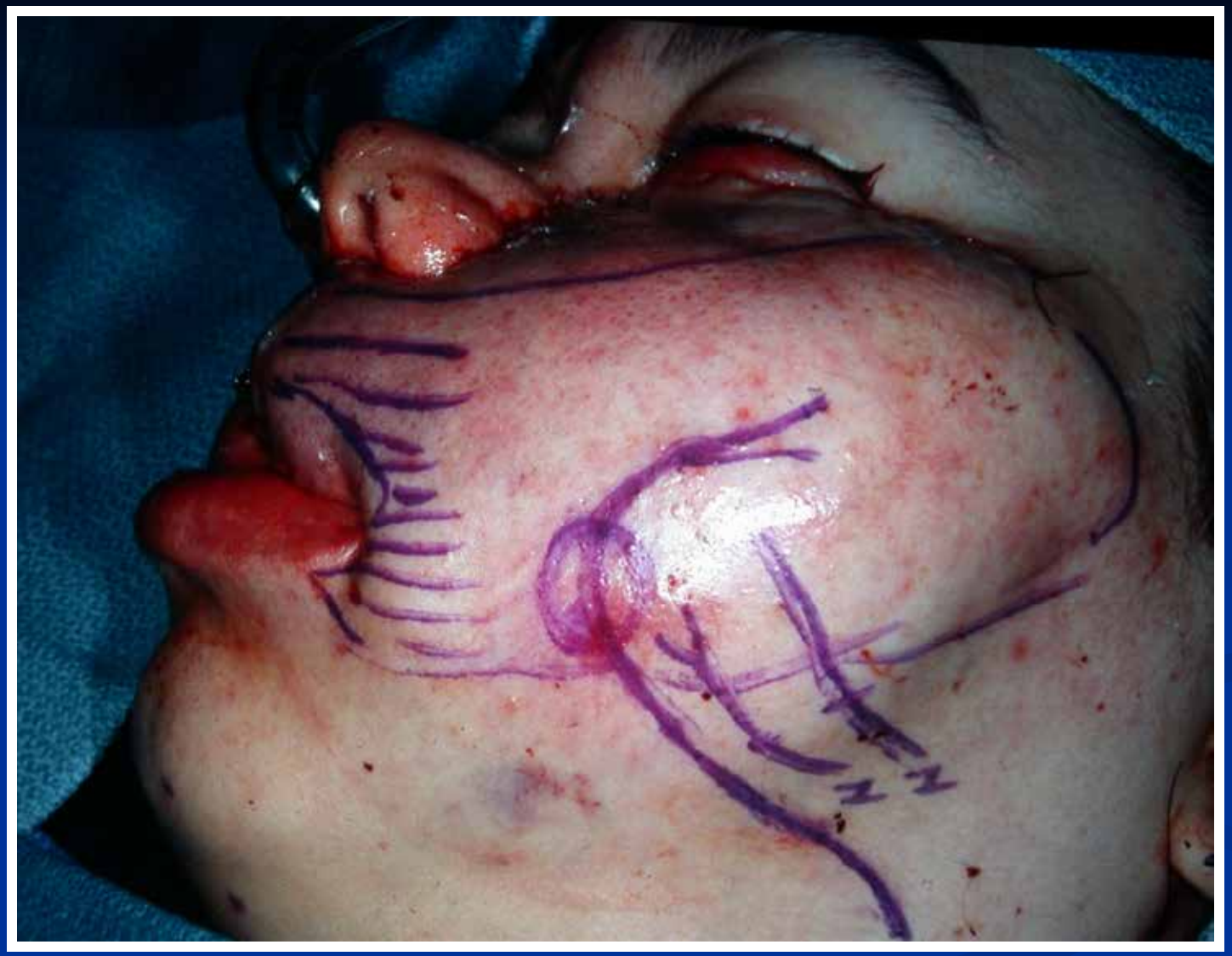


Tailored functional Rectus Abd. M/C flap

Post flap transfer



Intercostal nerve to facial nerve repair (x2)

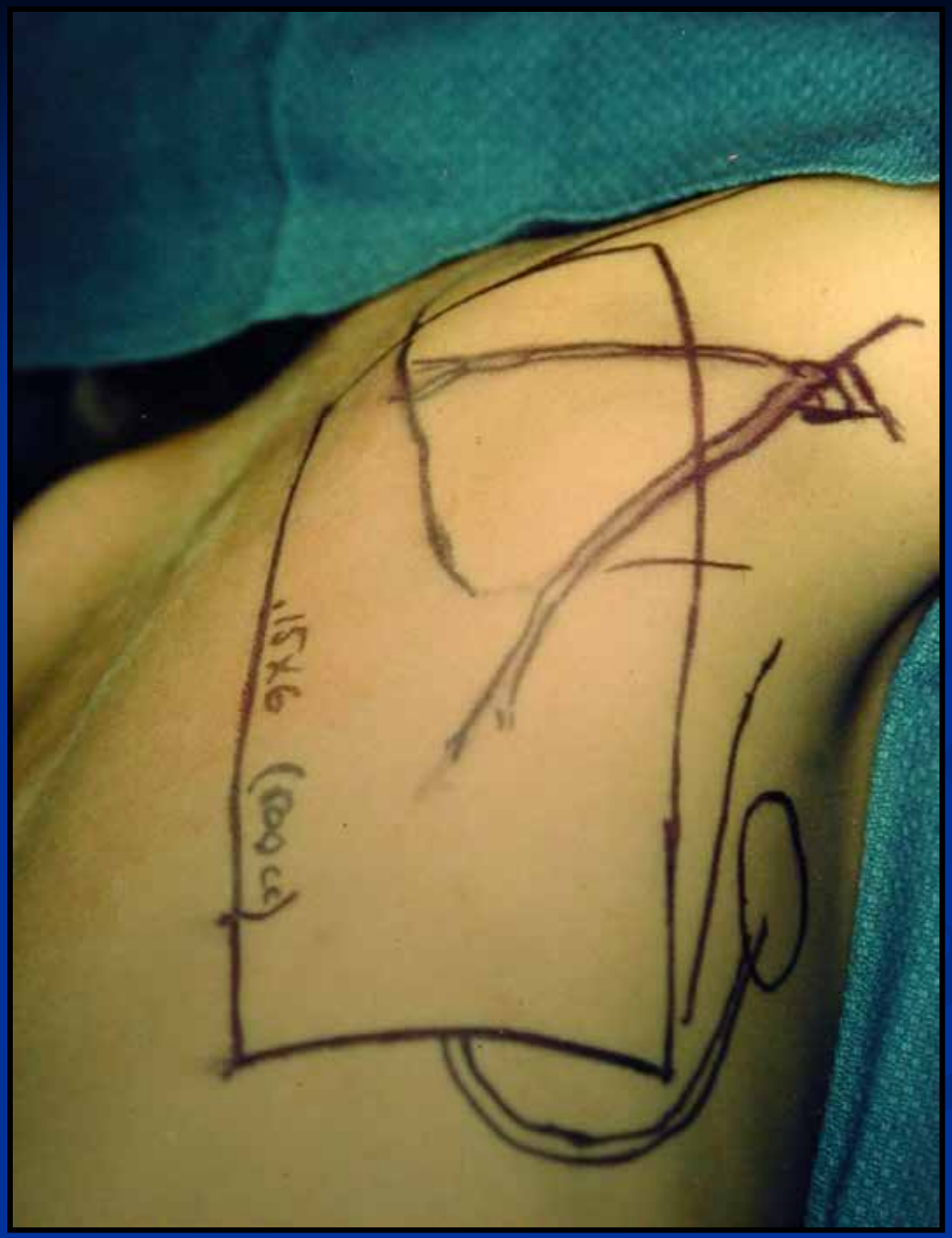


Flap transferred – functional rectus abdominus

nerves coapted to facial br

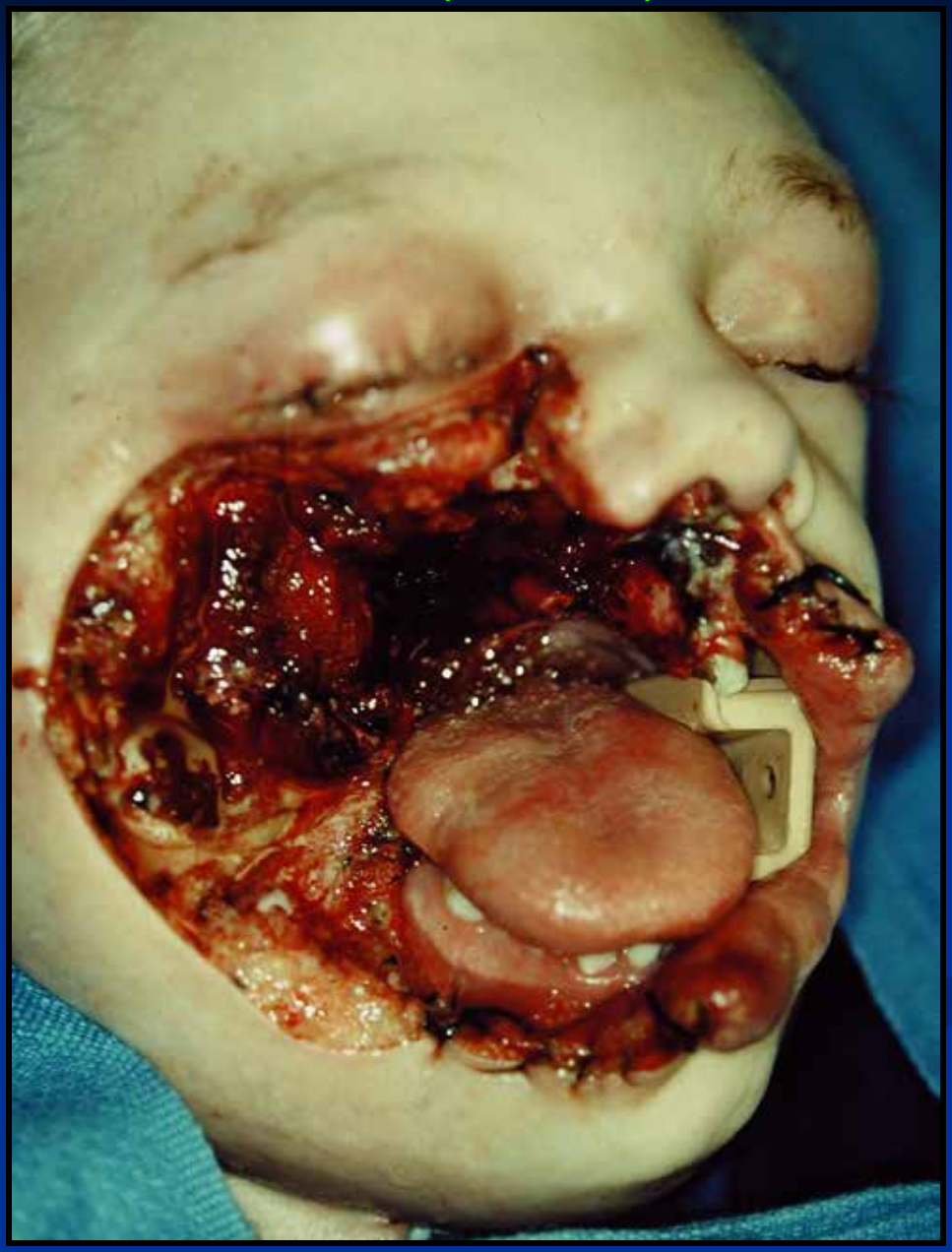
5F – extensive AVM of (R) cheek, maxilla, upper & lower lips

Preoperative expansion of back for composite folded flap transfer



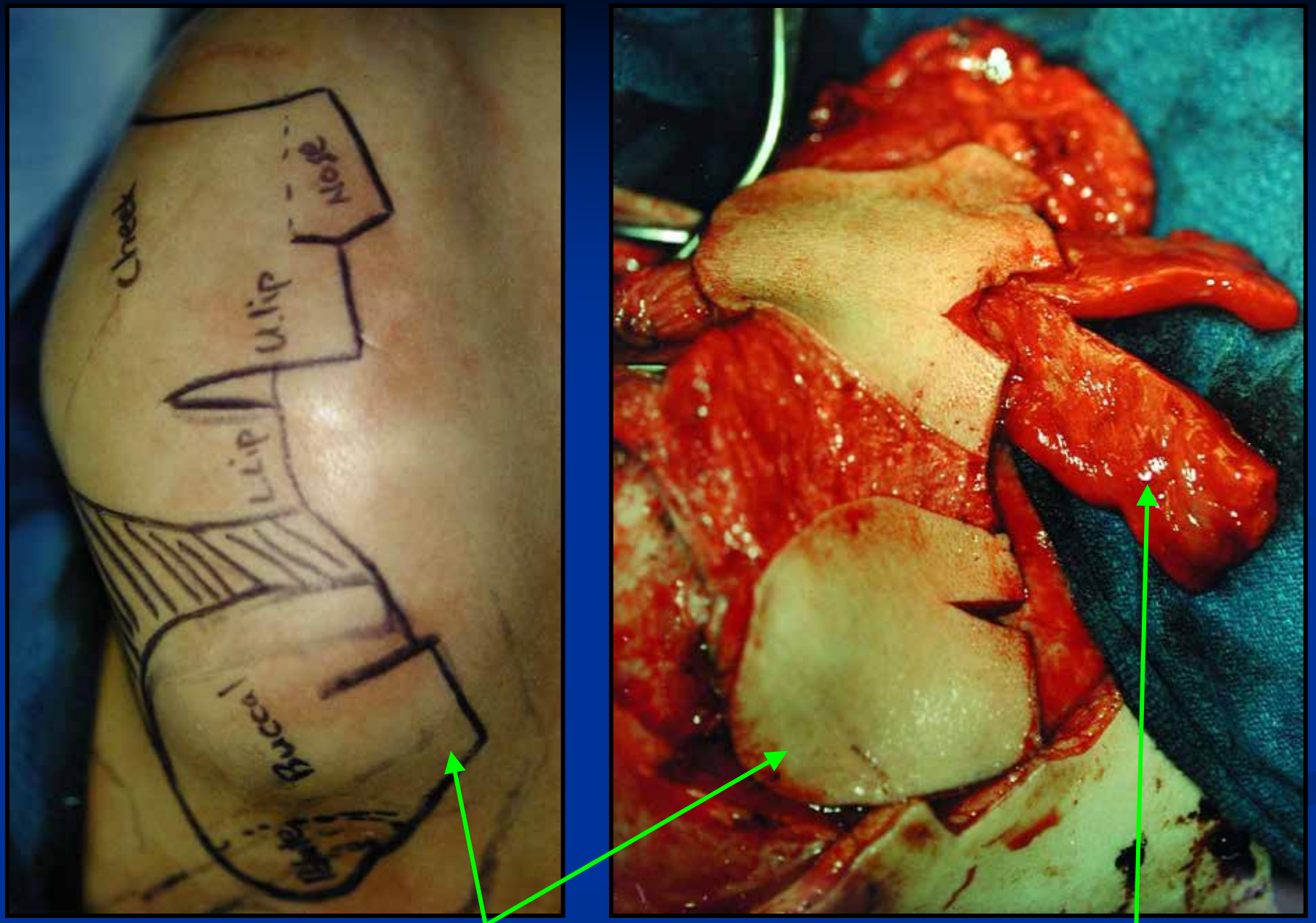
Post AVM resection -massive blood loss
-wound packed

5 days post resection
- Contaminated (? infected) wound



Alginate model of defect

3-D model g 2-D pattern



Tailored expanded parascapular flap & lat. functional lat. dorsi muscle

Folded parascapular/lat dorsi sandwich flap in situ

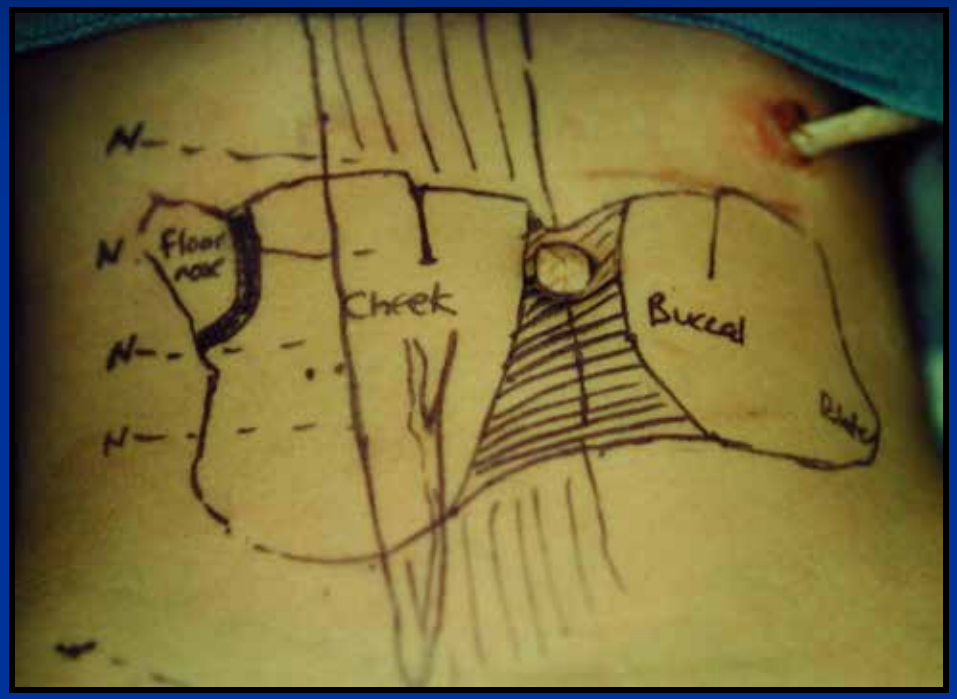
prior to detachment

Post flap transfer

4 days post op

- total flap necrosis due to infection

Post debridement & SSG



Tailored functional TRAM folded flap

Post flap transfer

2 years post op

4 years post op

6 years later

- Expanded submental flap & cheek flap for cutaneous coverage

39M extensive AVM (L) cheek, lips, maxilla

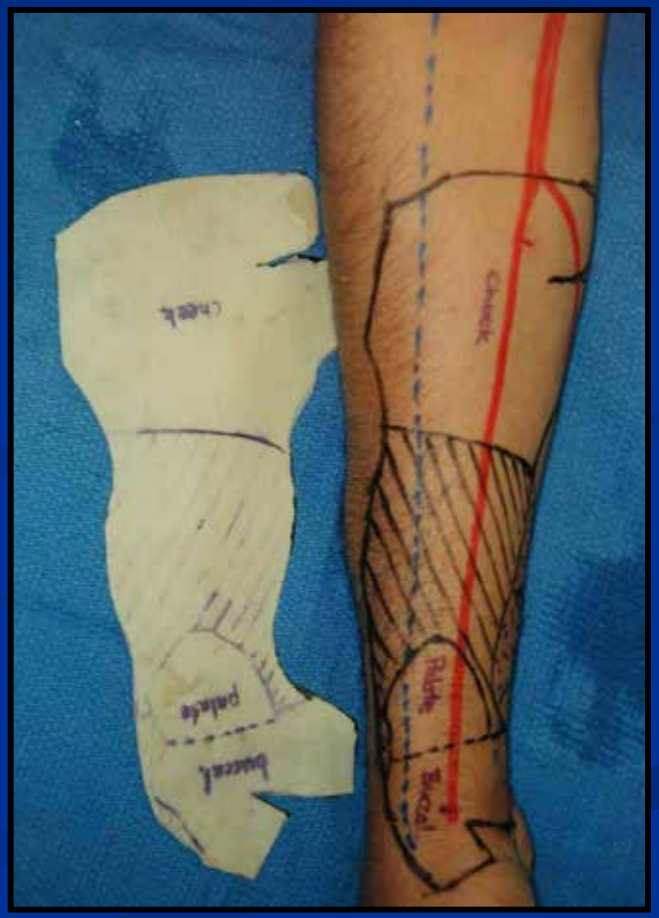
Alginate model of defect

3-D MODEL → 2-D PATTERN

VOLUME

COVER/LINING (S)

3-D model g 2-D pattern

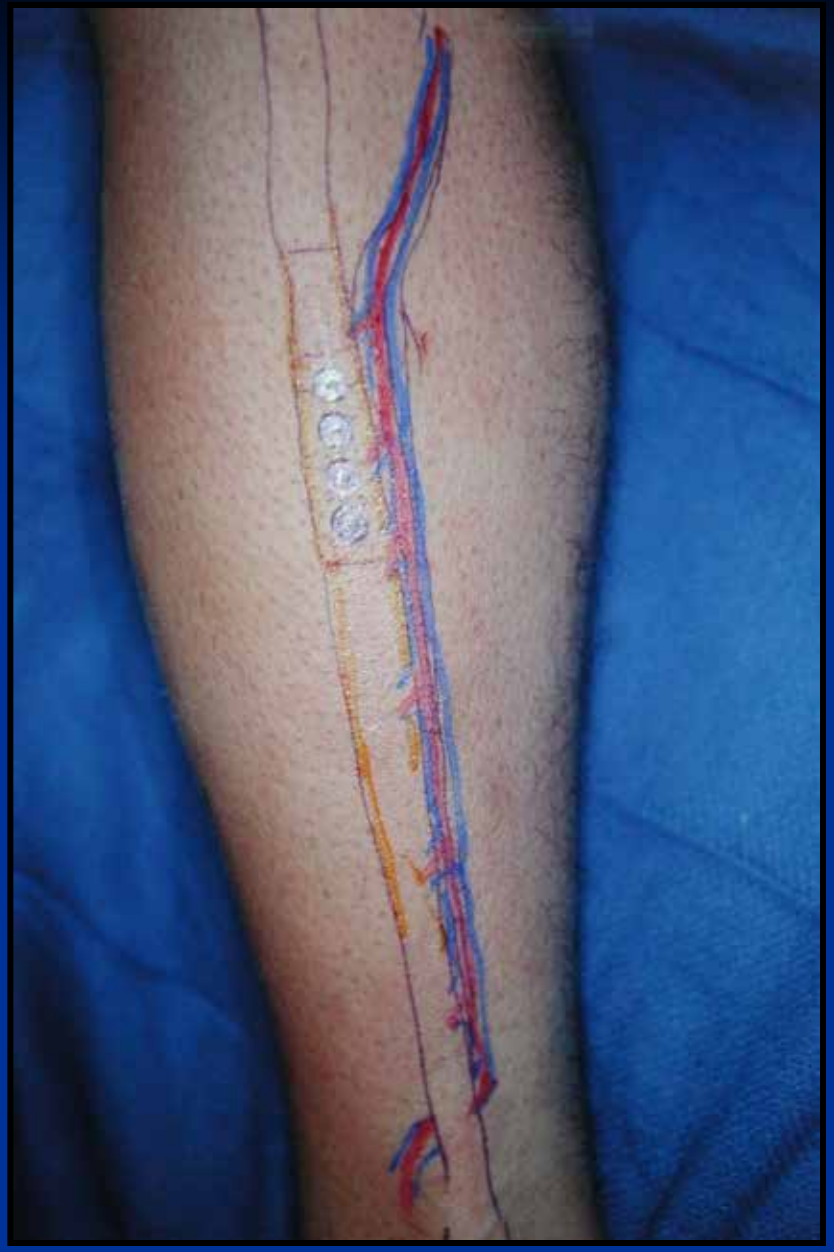


Tailored radial forearm flap

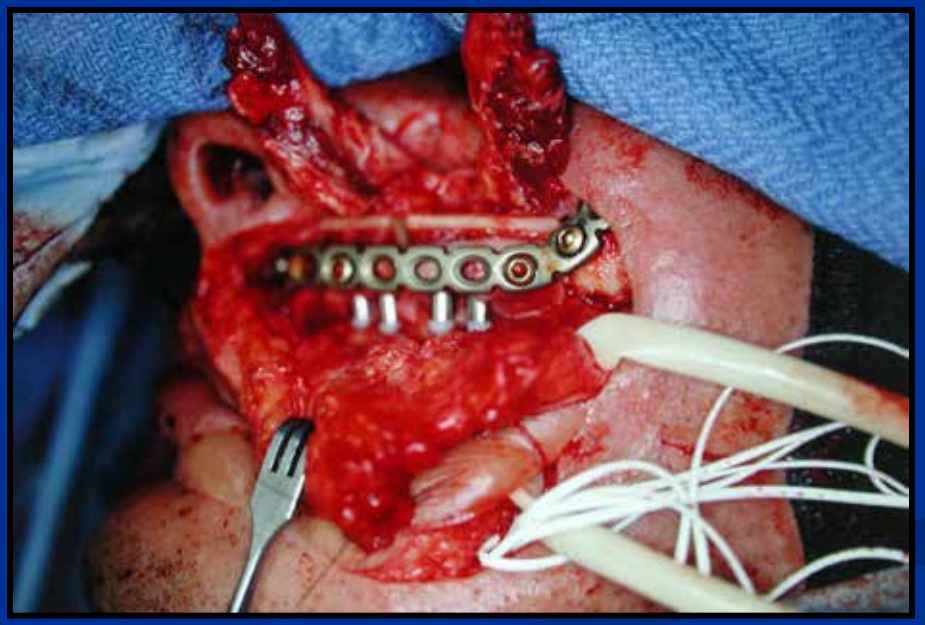
Contralateral FAMM flap for vermillion

& palmaris longus oral sling

Immediate post op



Free fibula flap with osseointegrated implants for (L) maxilla reconstruction

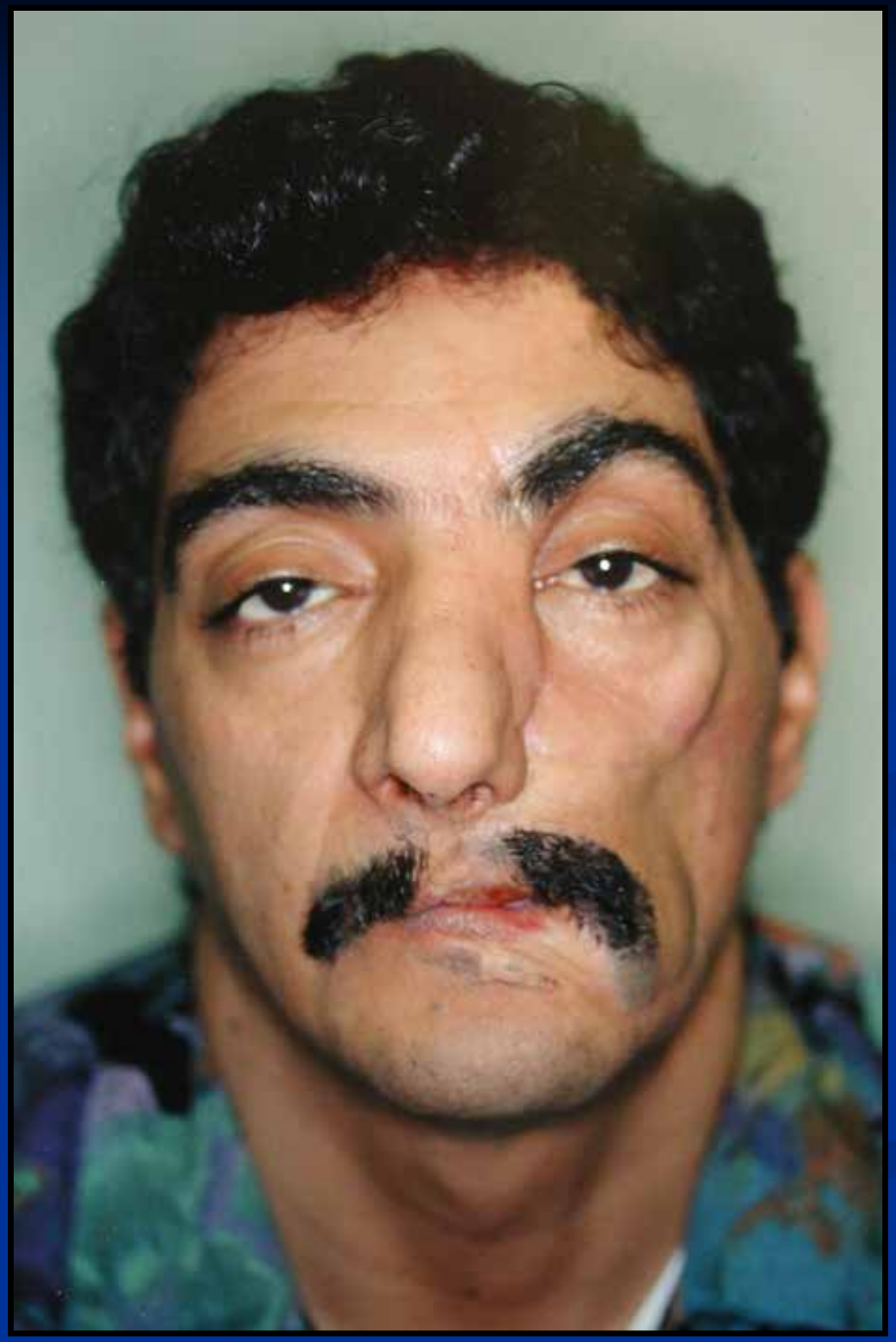


Free fibula flap with osseointegrated implants transferred to (L) maxilla

(Willie Stephens)

Extended forehead flap to cover cheek defect & moustache reconstruction

6 months post op



Functional submental/platysmal flap for cheek reconstruction

Functional submental / platysmal flap based on submental vessels & cervical branches of facial N.

1 week later

- muscle functioning

2 years later

AVM Defects

- Reconstruction of AVM defects of central face with **Prefabricated** & **Prelaminated** Flaps

Flap Prefabrication

Vascular Pedicle

–

Subcutaneous Buried Position

–

Neovascular Response

–

Flap Transfer

Flap Prelamination

Implantation of tissue or other device into a vascular territory prior to its transfer

(Vascular supply is not manipulated)

Massive full thickness necrosis of the (R) cheek, upper & lower lips.

Post embolization of vascular malformation

Thrombosis of multiple

ext carotid branches

Residual deformity post multiple tissue expanders & free radial forearm flap for (R) cheek lining

Plan for prefabricated neck flap for lower lip reconstruction

Free mini DLFC vascular pedicle flap to neck

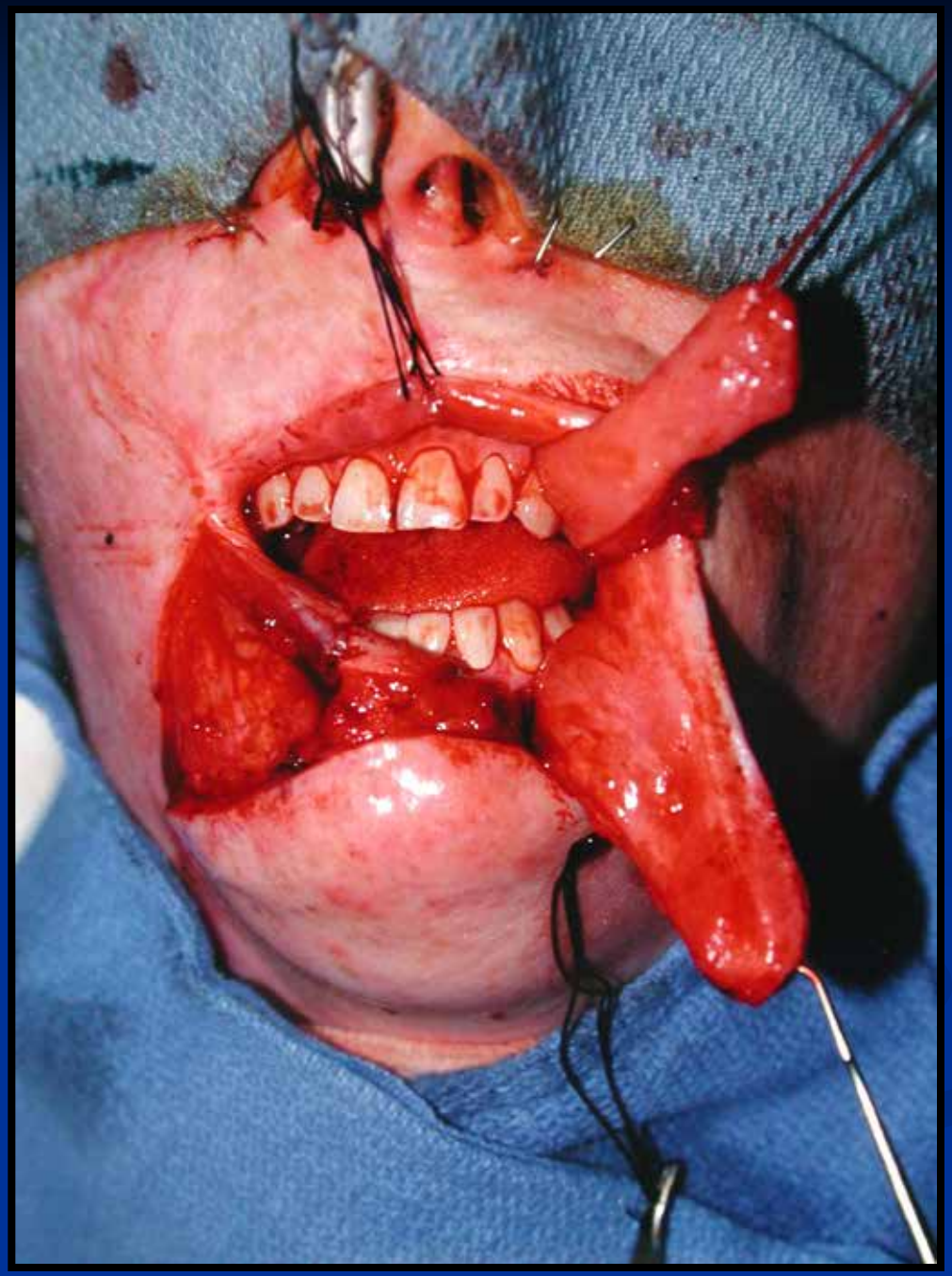
Placed over a tissue exp

8 weeks later

Lining flaps for lower lip

&

FAMM flap for upper lip

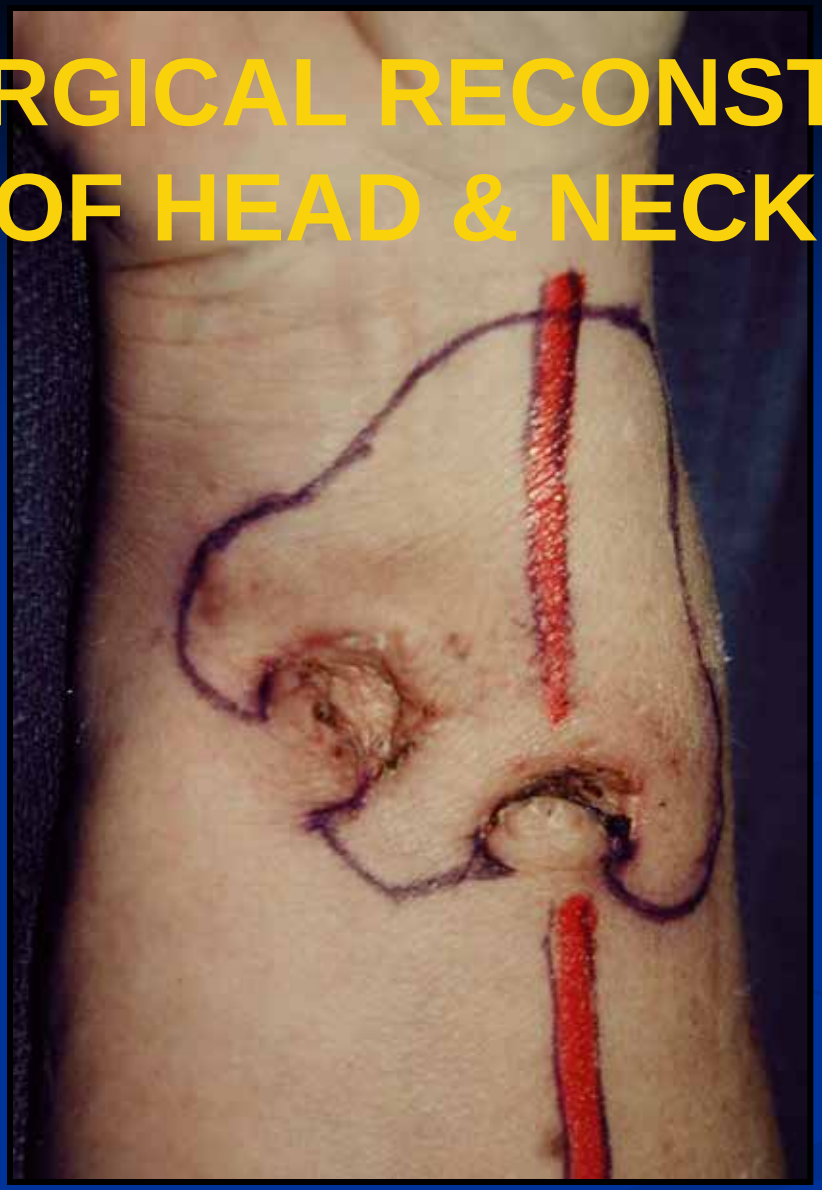


Immediate

Prefabricated flap is slightly congested – chemically leeching

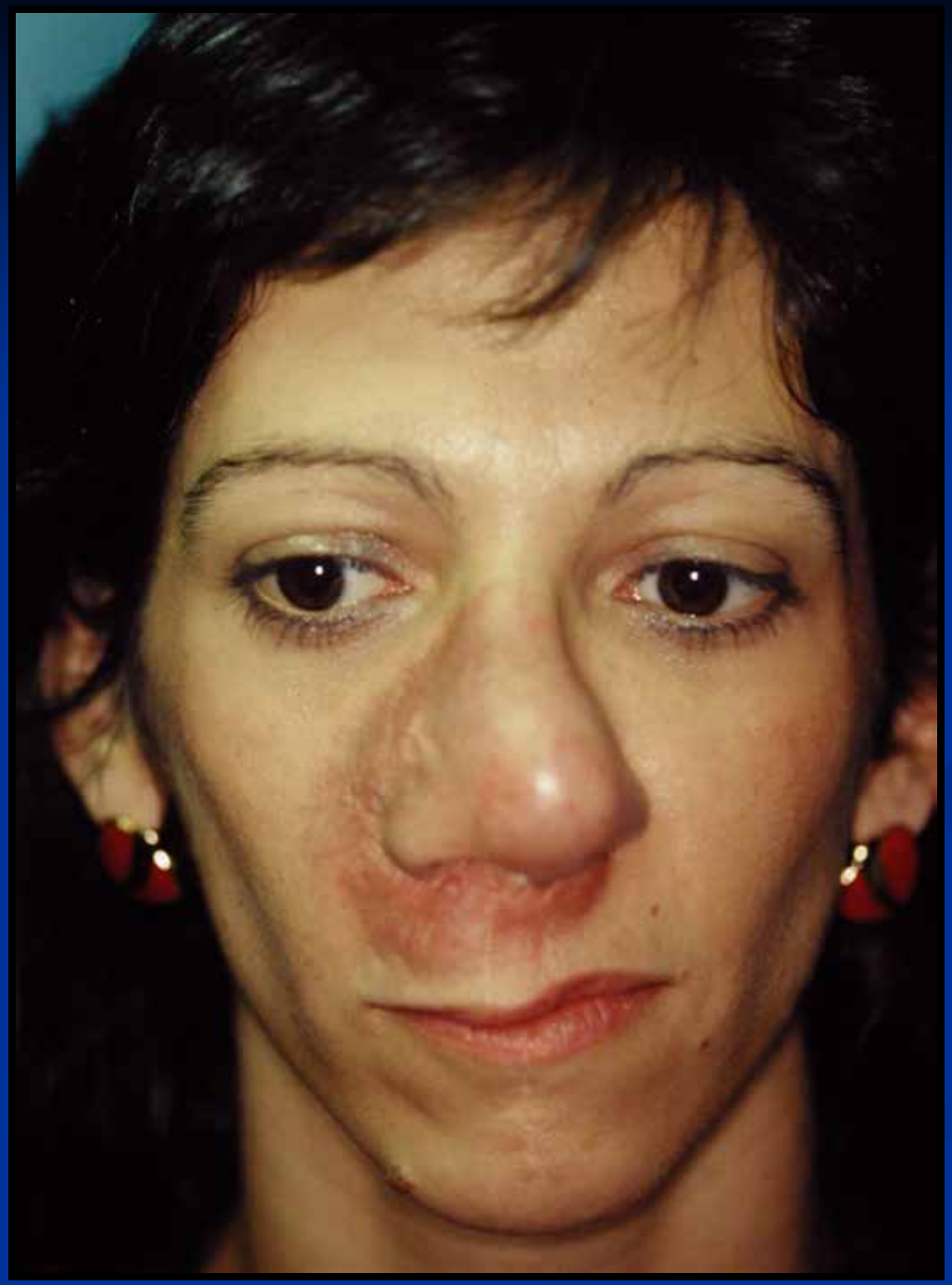
6 months

MICROSURGICAL RECONSTRUCTION OF HEAD & NECK



Multifaceted & Laminated Flaps

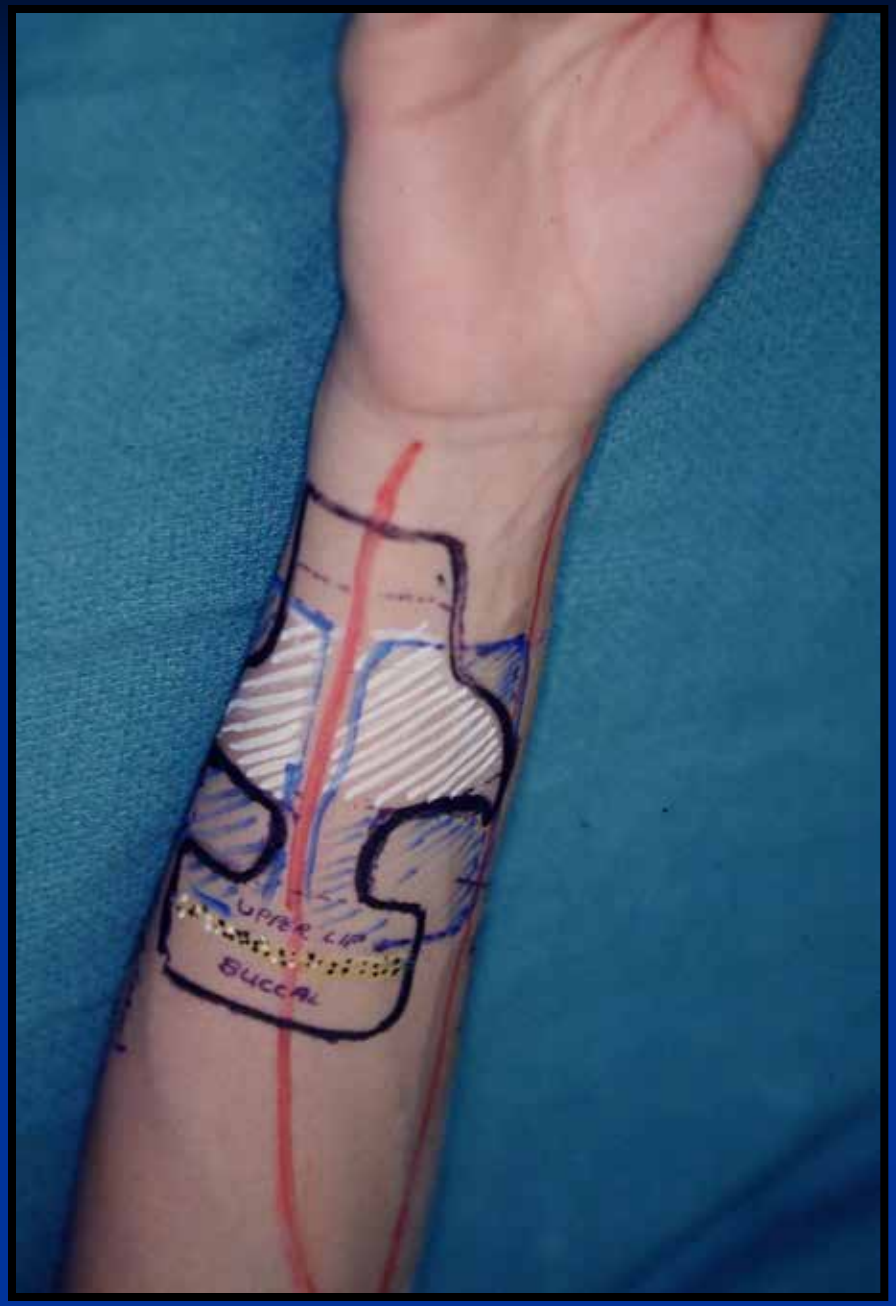
Prelaminated

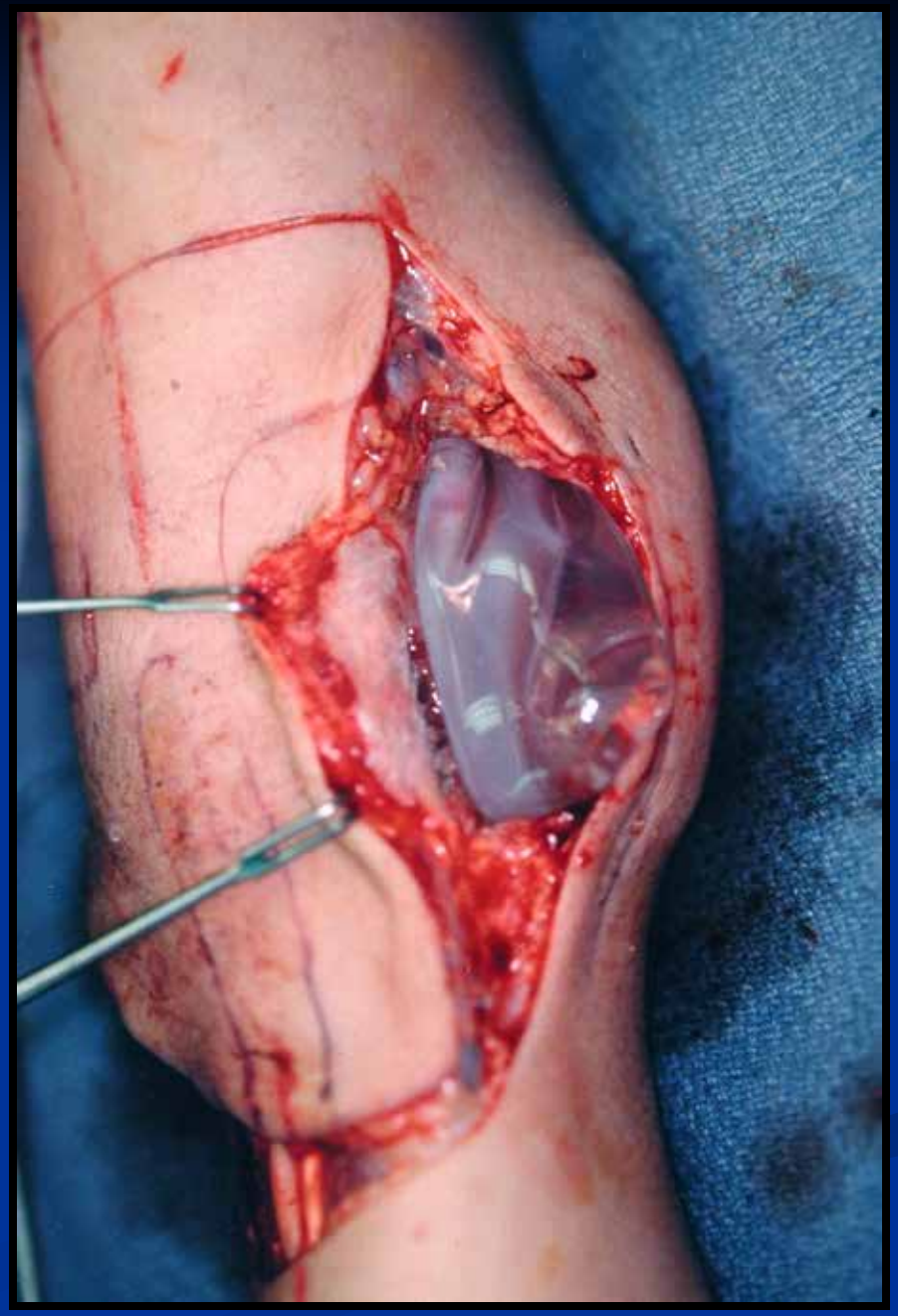


Young Patient with AVM of nose/lip

(prior partial excision & Abbe flap)

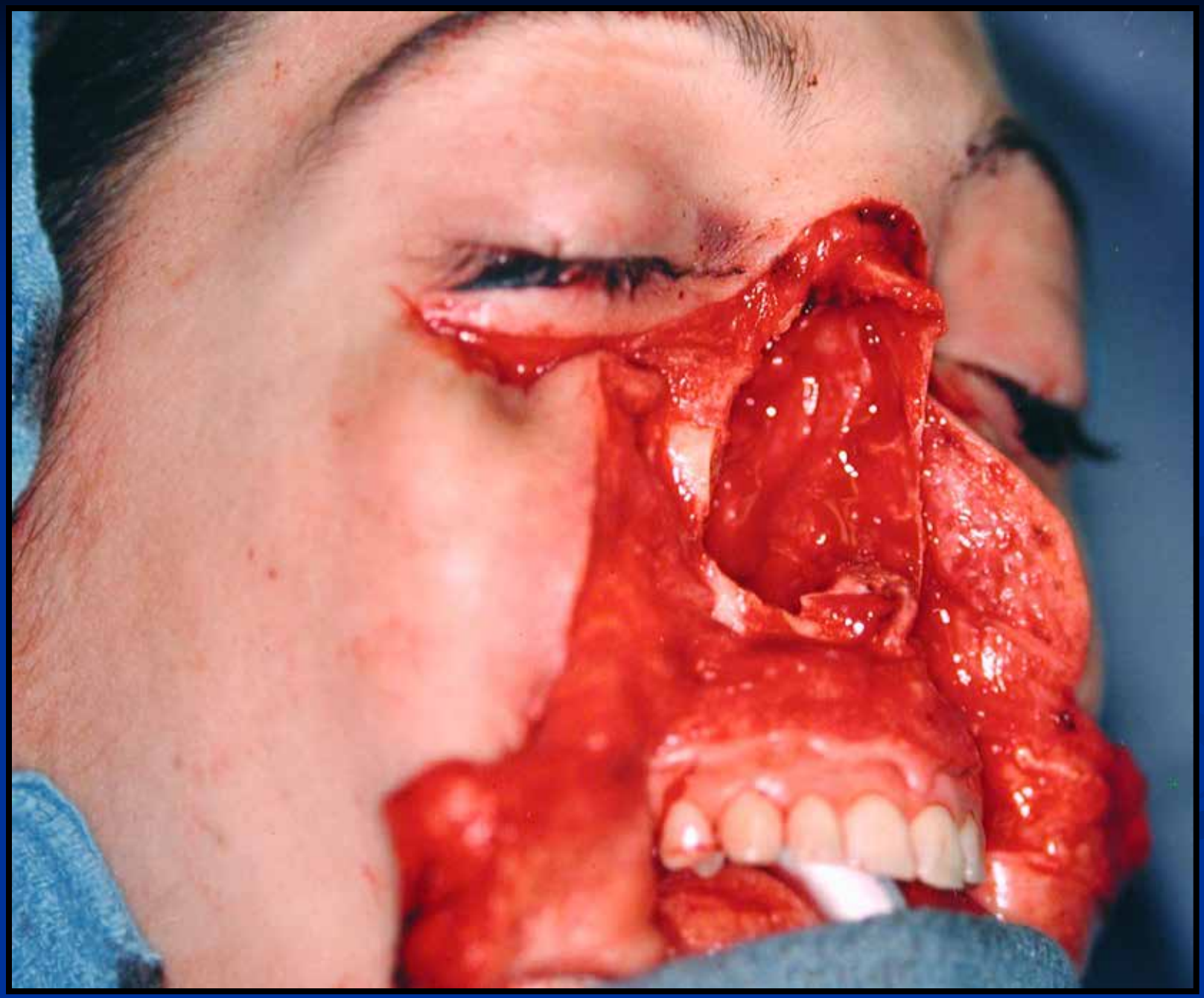
Prelaminated ulna forearm flap





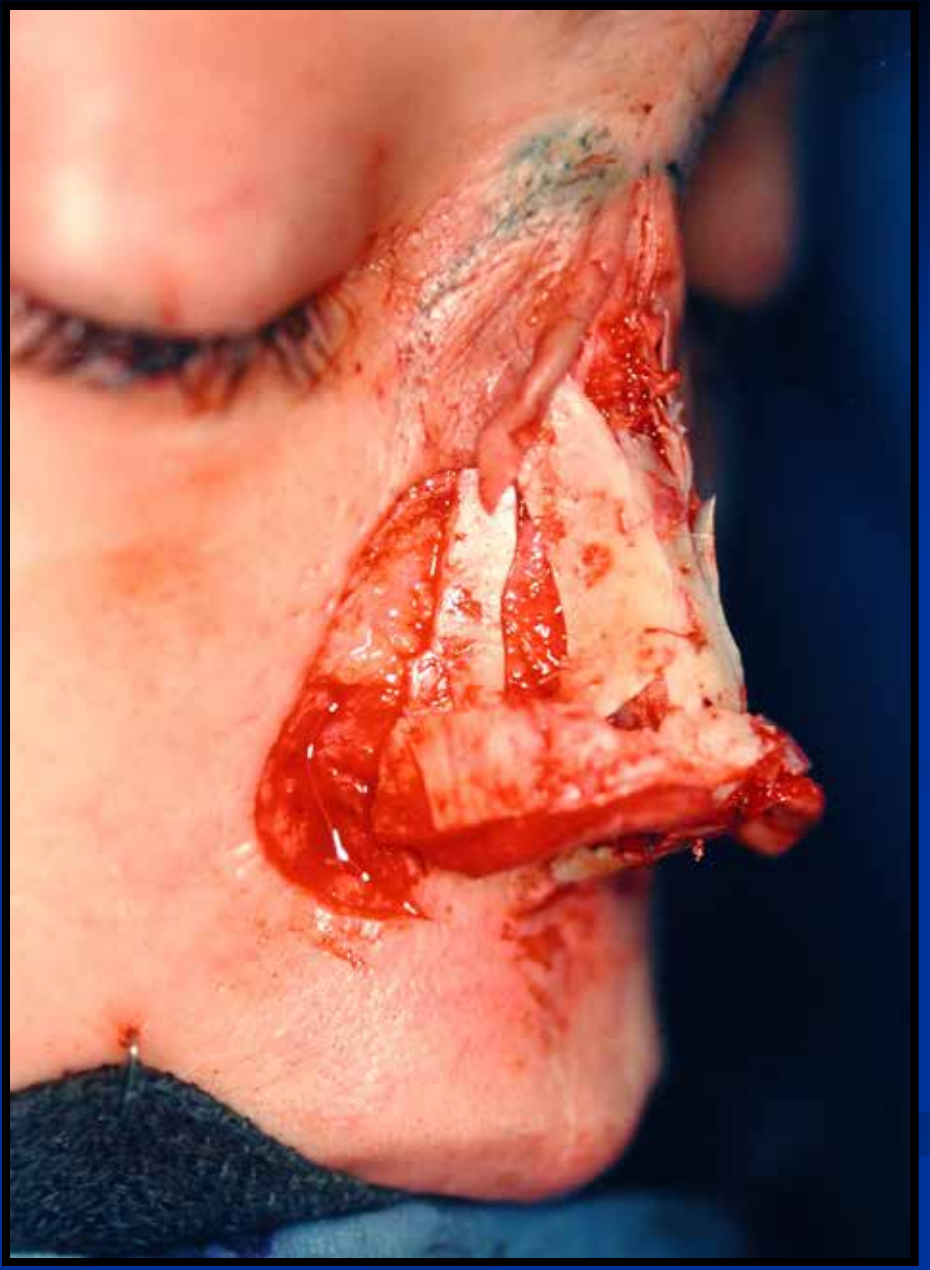
Healed FTSG on forearm flap

Resection of AVM



Prelaminated flap transfer for total nasal & upper lip reconstr.

Post first revision

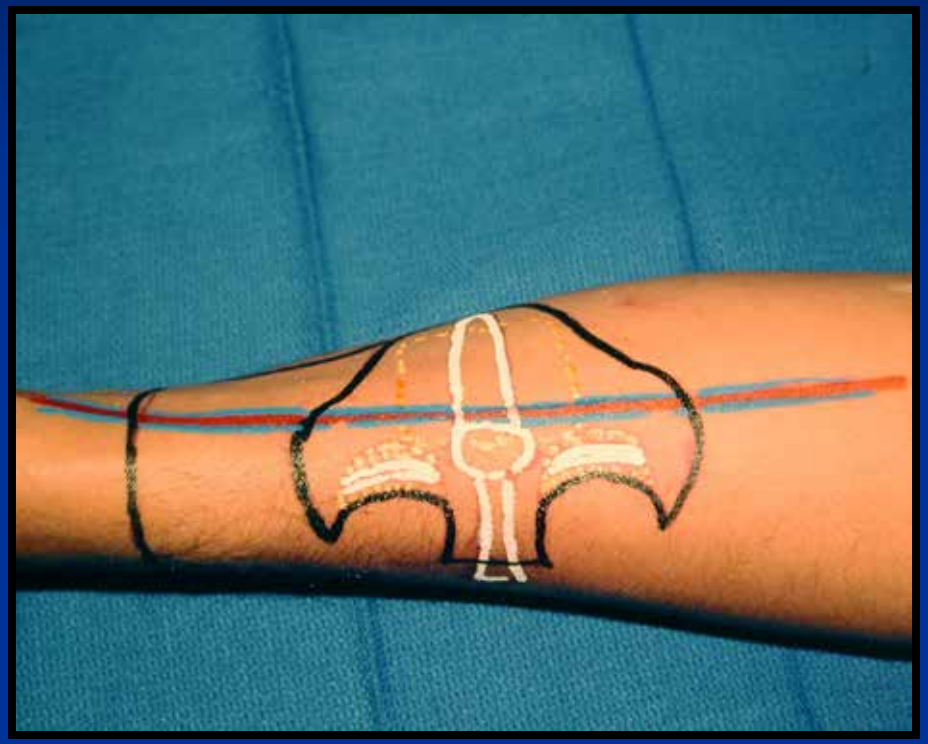


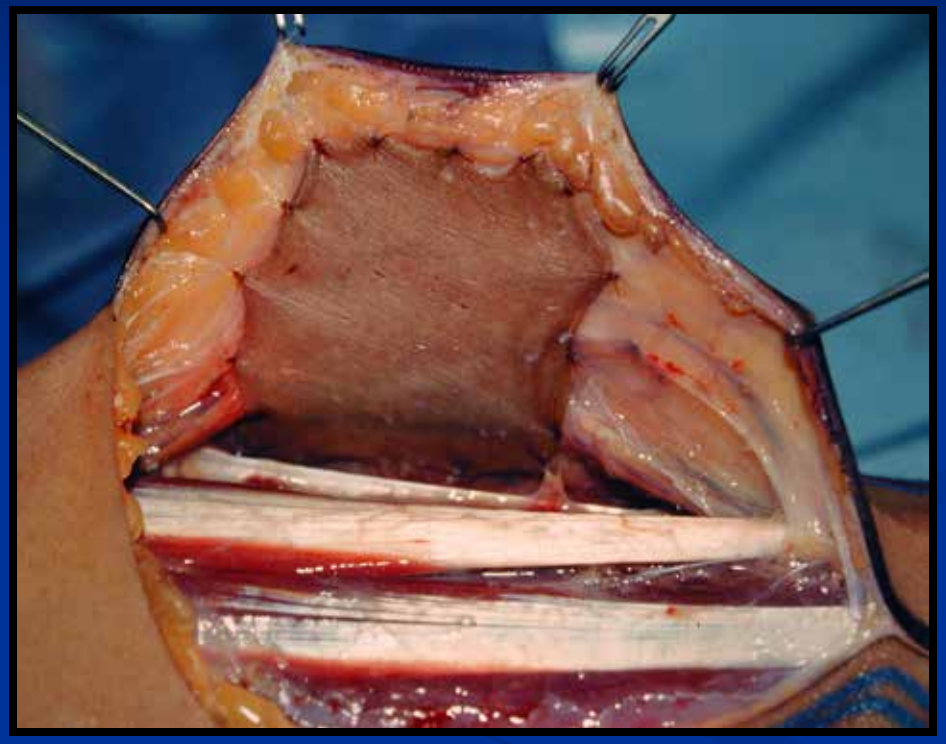
Second stage nasal reconstruction with lining flaps, cartilage grafts & forehead flap

12 month post op

12 month post op

AVM of nose & cheek



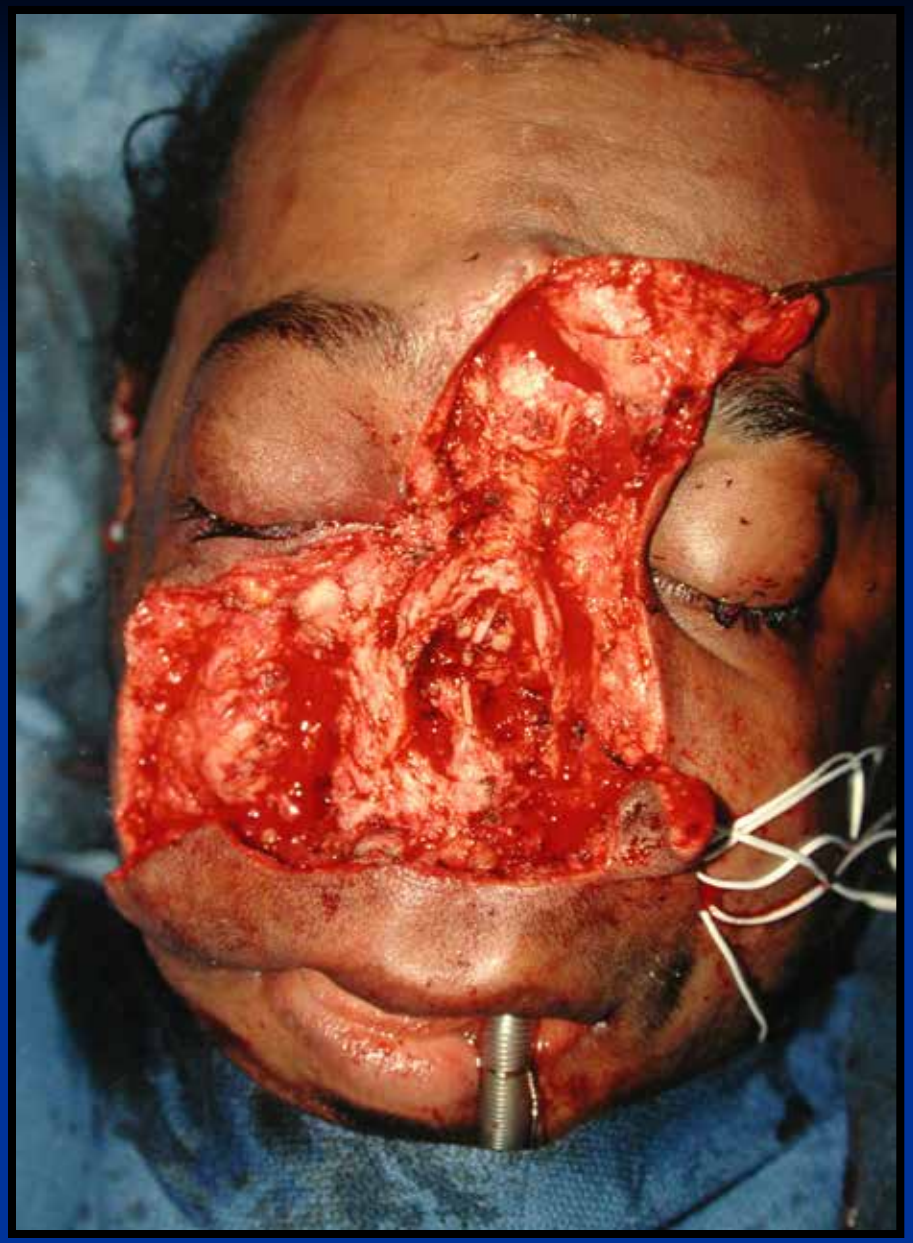


Prelaminated flap on forearm for nasal & cheek reconstruction

Carved costal cartilage graft for support

FTSG for lining

Prelaminated nose & cheek flap on forearm



Post resection AVM

Early post op

Post cheek advancement

Final coverage with expanded forehead flap

AVM of nose & cheek

AVM Head and Neck
Free Flap Reconstruction

- Allows wide resection
- Optimizes vascularity :

  (? less ischemic stimulus)
- ? residual AVM adj to flap can remain quiescent

Fate of Residual AVM Following Treatment

- Not known
- Free flap brings in new vascularity and nutritional blood flow
  - ? Residual AVM becomes asymptomatic

Conclusion - I

1. Wide surgical excision and complex reconstruction “cures” approx 50% of patients.
2. All patients (even residual AVM) greatly improved symptomatically.
3. ?? Can residual AVM “invade” FF??

   -probably can & will if high flow AVM left at base of excision

Conclusion - II

4. Need better way to determine adequacy of resection.
5. Consider earlier resection prior to symptoms.
6. Interdisciplinary/interinstitutional efforts to improve understanding of etiopathogenesis (molecular genetics) and treatment of AVMs.