Juvenile Nasopharyngeal Angiofibroma

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JNA

- History
- Definition
- Route of spread
- Staging
- Surgical approach
- Conclusion

History

- Hippocrates in 5th century : Description of nasal polyp
- Chelius 1847: Describe fibrous nasal polyp association with puberty
- Friedberg : coined the term as Angiofibroma
- Radkowski 1996: gave classification of juvenile nasopharyngeal Angiofibroma

Disclaimer

- The author has taken some of the pictures from various sources purely because of academic purpose. However the study is conducted by me during the year 2002 to 2010

Definition

- Juvenile Nasopharyngeal Angiofibromas are Benign, locally aggressive, highly vascular, non encapsulated tumors.
- Commonly affecting predominantly adolescent males.
- These lesions though benign histologically yet they may become life-threatening with excessive bleeding or intracranial extension.

Occurrence

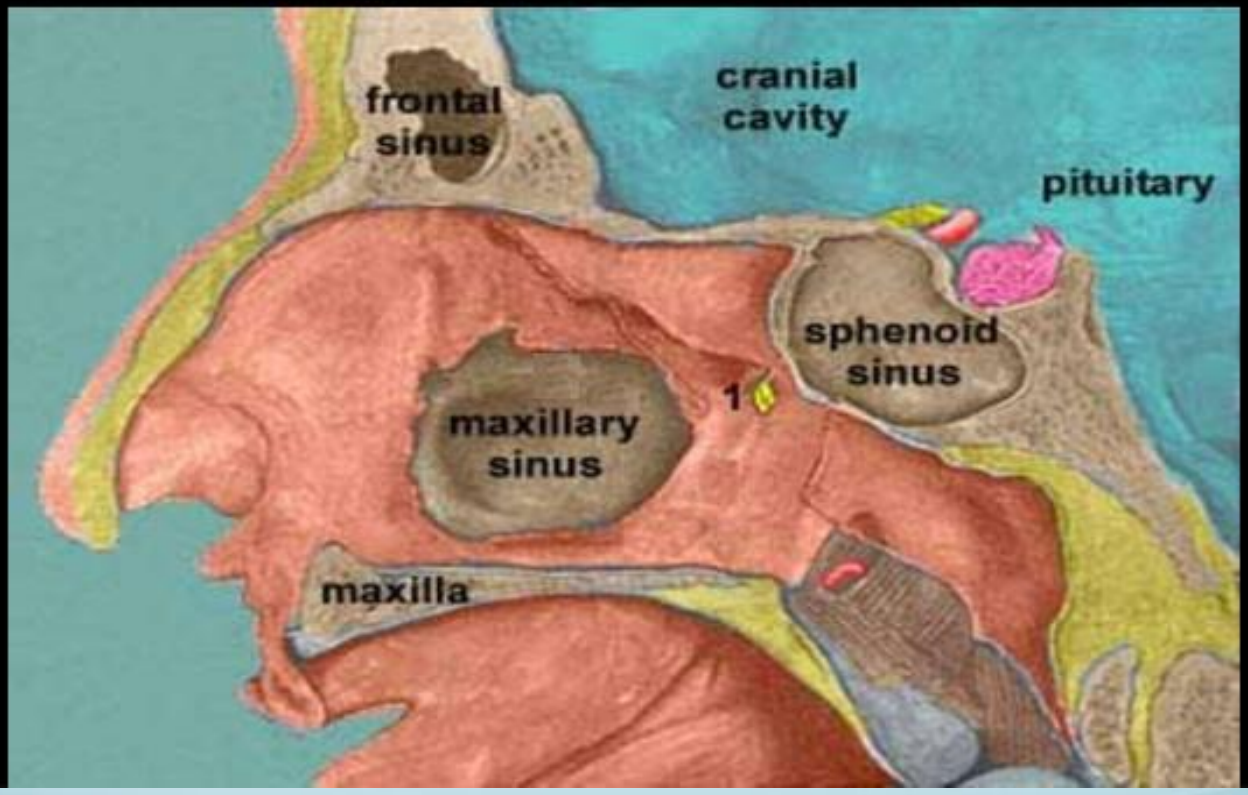
- JNA accounts for 0.5 % of all head and neck tumors
- Occurring almost exclusively in males but also reported in children, elderly, young and even pregnant women
- Average age of onset is around 15 years.

Pathogenesis

- Tumor consists of proliferating, irregular vascular channels within a fibrous stroma
- Tumor blood vessels typically lack smooth muscle and elastic fibers contributing for sustained bleeding (Tunica media)
- Stromal compartment is made up of plump cells, they give rise to certain amounts of collagen resulting in varying degree of consistency which may be hard firm or relatively soft.
- Androgen receptors are present in at least 75 percent cases
- Much smaller proportion have some progesterone receptors
- No estrogen receptors have been demonstrated.

Site of origin

Arises in posterior nasal cavity, near superior border of sphenopalatine foramen



Appearance

- Smooth lobulated
- Pale or purplish
- Reddish grey
- Beef red
- Compressible



© Hodder Arnold / Scott-Brown 7E

Diagnosis

- History
- Physical examination
- Radiology
- CT scan
- MRI
- Angiogram

History (Contd)

A. **Major Complaints:**
   - Progressive nasal obstruction.
   - Recurrent severe epistaxis
   - Headache
   - Hyposmia/anosmia

B. **Minor Complaints**
   - Facial swelling
   - Facial pain
   - Hearing loss
   - Nasal intonation to voice
   - Trismus
   - Diplopia
   - Proptosis
   - Visual loss

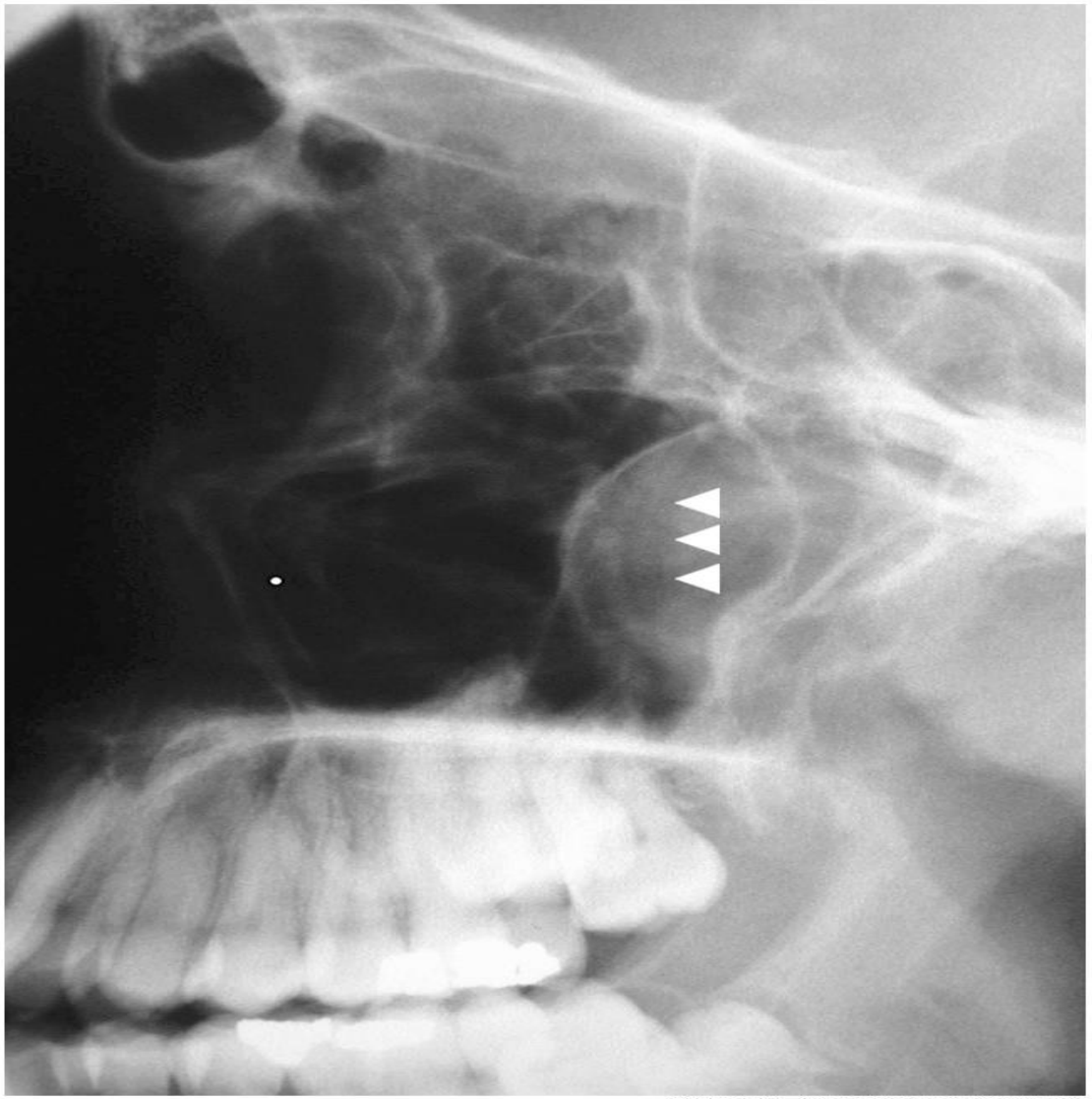
Examination

- Abundant mucopurulent secretion in nasal cavity.
- Pink or reddish mass in nasopharynx.
- Soft palate displaced inferiorly.

Investigations

- **CBC** → Hyochromic microcytic anaemia due to chronic blood loss.
- **Plain lateral skull radiographic**: → Anterior bowing of posterior wall of maxillary sinus.
- **Combined CT scan & MRI** to determine exact extent or stage of the tumour.
- **Diagnostic angiography** is undertaken to evaluate source of blood supply & as a prelude to selective embolisation
- **Trans-nasal biopsy** is not necessary and can provoke haemorrhage, usually not done in our settings

Holman Miller Sign



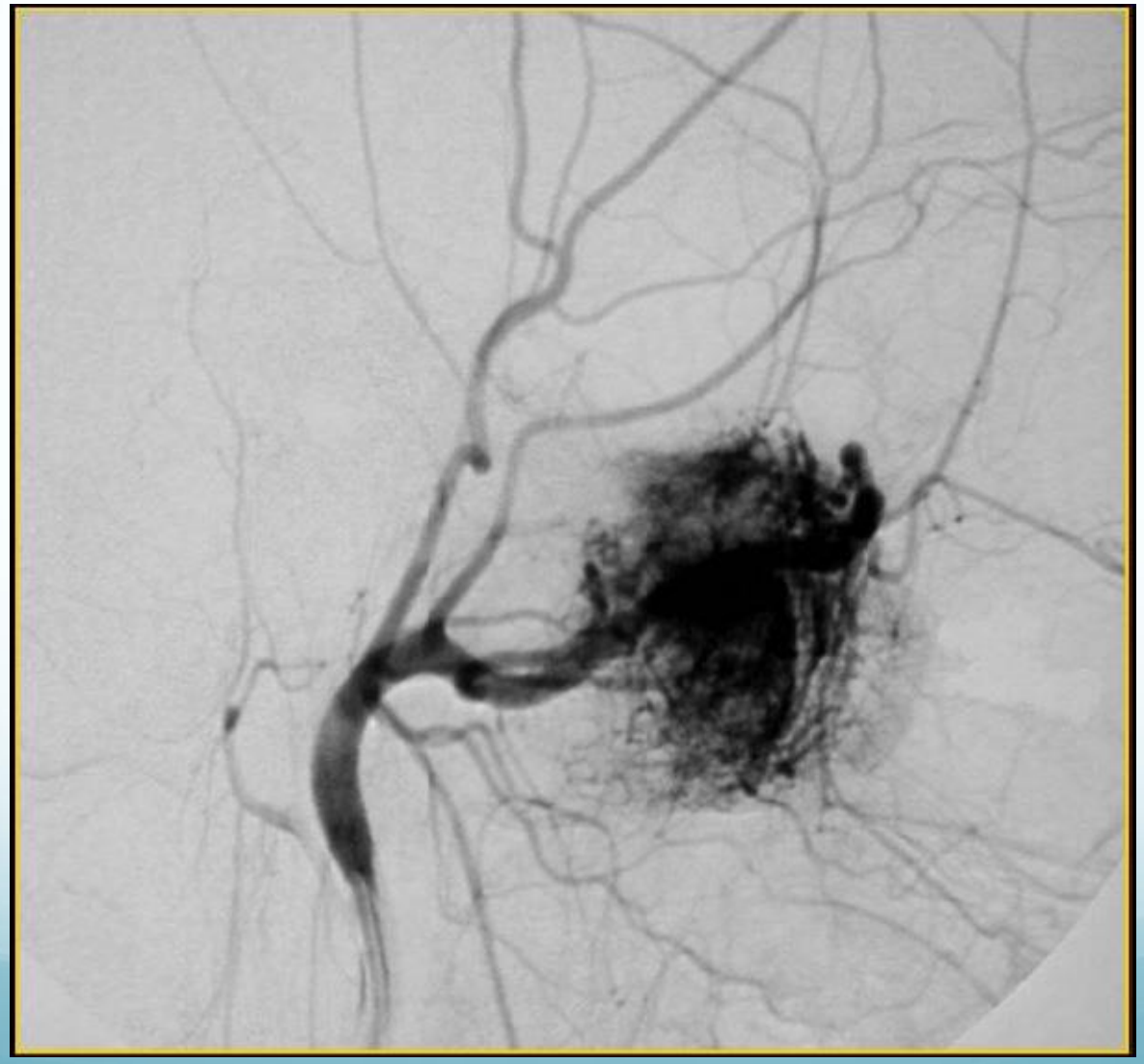
CT Scan Axial View



CT Scan Coronal View



Angiography (pre-embolised)



Angiography (Post-embolisation)



Intracranial Extension

- 20-36 % intracranial extension.

- Intracranial extension is almost always extramural (Scott Brown)

Route of spread

- Medially: Nasal cavity & nasopharynx.
- Laterally: Pterygopalatine fossa & infratemporal fossa.
- Superiorly: Orbit, middle cranial fossa, cavernous sinus & pituitary gland.
- Inferiorly: oral cavity

Classification/Staging

- Sessions classification (1981)
- Fisch classification (1983)
- Chandlers classification (1984)
- Modified Fisch classification (Andrews; 1989)
- Radkowski classification (1996)
- University of Pittsburg Medical Center Staging (2008)

Fisch Staging System

- **Stage I** Tumour limited to nasopharyngeal cavity; Bone destruction negligible or limited to sphenopalatine foramen
- **Stage II** Tumour invading pterygopalatine fossa or maxillary, ethmoid or sphenoid sinus with bone destruction
- **Stage III** Tumour invading infratemporal fossa or orbital region
  A. Without intracranial involvement
  B. With intracranial extradural (parasellar) involvement
- **Stage IV** Intracranial intradural tumour:
  A. Without infiltration of cavernous sinus, pituitary fossa or optic chiasma
  B. With infiltration of the cavernous sinus, pituitary fossa or optic chiasm

Radkowski Classification of JNA

Stages

1A: limited to nose and nasopharynx

1B: Extension to at least one paranasal sinus

2A: Minimal extension through sphenopalaltine foramen, includes minimal part of pterygomaxillary fossa

2B: Full occupation of pterygomaxillary fossa with Holman-Miller sign, may have superior extension with orbital bone erosion

2C: Extension through pterygomaxillary fossa into cheek and temporal fossa

3A: Skull base erosion with minimal intracranial extension

3B: Skull base extension with extensive intracranial extension+ cacernous sinus

Treatment

- Surgery: Gold standard
- Radiotherapy: Unresectable life threatening
- Chemotherapy: Recurrence of previous surgery
- Hormonal: Diethylstilbestrol and flutamide



Figura 8. Peça cirúrgica.

Surgical Approaches

- Trans-palatal approach (Wilson)
- Sub-labial trans-palatal approach (Sardanas)
- Trans-maxillary
  - Extended lateral rhinotomy incision
  - Mid-facial degloving incision
  - Denker's extended caldwell luc incision
  - Le-forte type-1 osteotomy
- Infra-temporal approach (Fisch)
- Anterior sub-cranial approach
- Image-guided, endoscopic, laser assisted removal

- Most small tumours: Transpalatal approach

  Lateral rhinotomy approach

- All stage tumors Open approaches
- Fisch staging Endoscopic approach

Hormonal Treatment

Oestrogens:

- Delays surgery, Secondary feminization

Flutamide

- Nonsteroidal androgen receptor blocker
- Also used in Mx of prostatic cancer
- Tumour shrinkage of up to 7 to 44 %
- Side effects: Nausea, breast tenderness, gynaecomastia
- Effects were only temporary & disappeared completely at end of therapy
- Role in preoperative preparation of **very advanced tumours, Intracranial extension**

Complications of surgery

- Recurrence (25-40 %)
- Prolonged nasal crusting leading to ozaena
- Infra-orbital nerve sensory deficits
- Nasal vestibular stenosis
- Displacement of globe
- Ophthalmoplegia
- Visual loss

Complications of Radiotherapy

- Growth retardation
- Pan-hypopituitarism
- Temporal lobe necrosis
- Cataracts
- Radiation keratopathy
- Skin, thyroid and nasopharyngeal malignancies

Follow Up

- Prolonged clinical and radiological monitoring is necessary for all patients due to high recurrence rate
- Disease-free status five years after primary surgery probably represents cure.

Recurrence

- Over expression of IGFII might be associated with a tendency to recurrence and poorer prognosis
- Juvenile Angiofibromas have also been reported to develop 25 times more frequently in patients with familial adenomatous polyposis (Scott Brown)
- Operative conditions after embolization are improved might influence adequacy of resection and reduce the rate of recurrence

Study

Study of 35 cases

Materials and Methods

- **Study Design**: Retrospective study
- **Study Setting**: Civil hospital

  Park Lane hospital
- **Study Duration**: May 2000 to 2010
- **Sample Size**: 35 patients

Eligibility Criteria

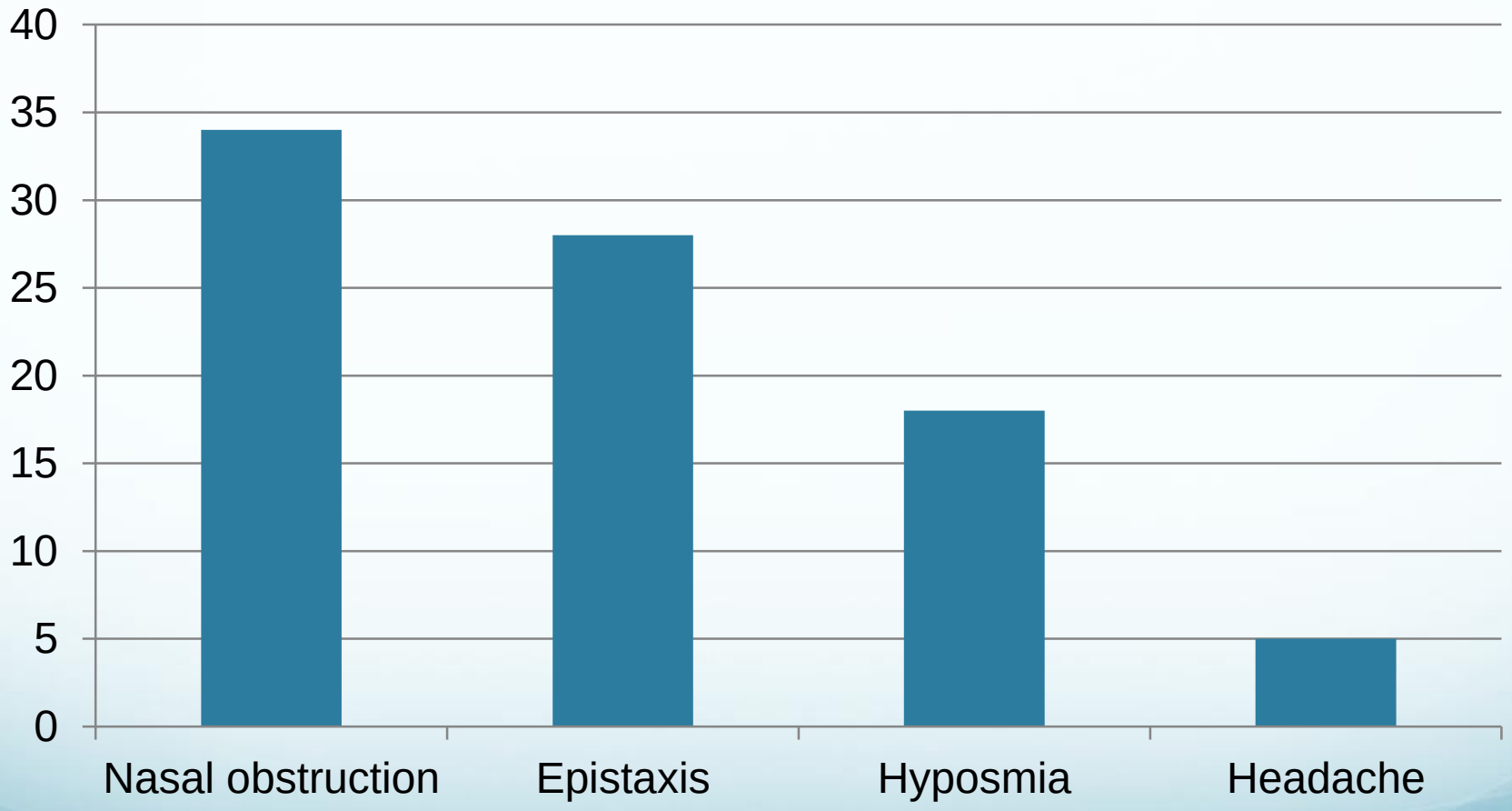
- **Inclusion Criteria:**
  - All cases diagnosed as Angiofibroma with and without intracranial extension
- **Exclusion Criteria:**
  - Unfit for surgery
  - Bleeding disorders

Results

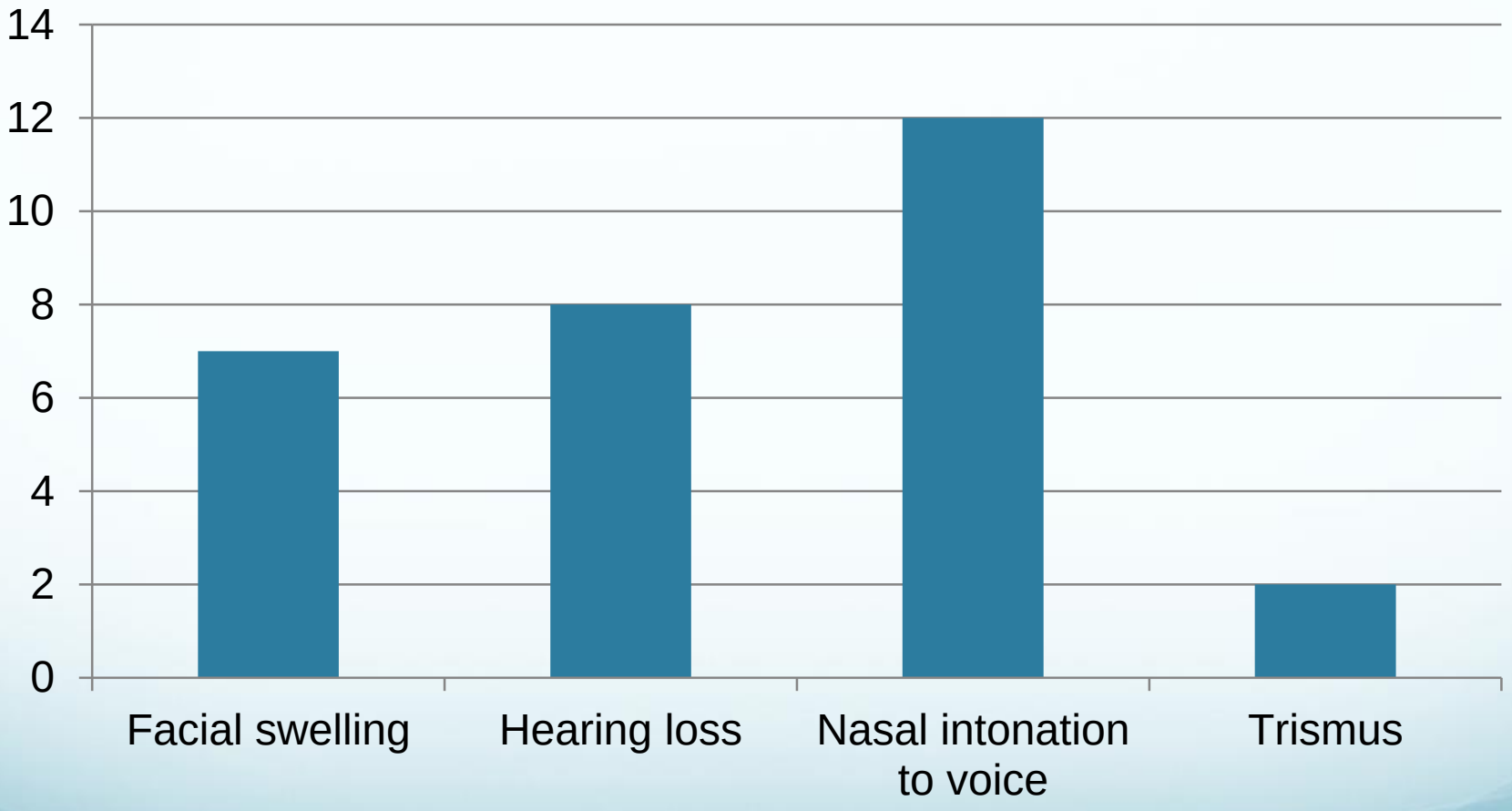
Age

Age	No. of patients
12-14	8
15-17	24
18-20	2
21-22	1

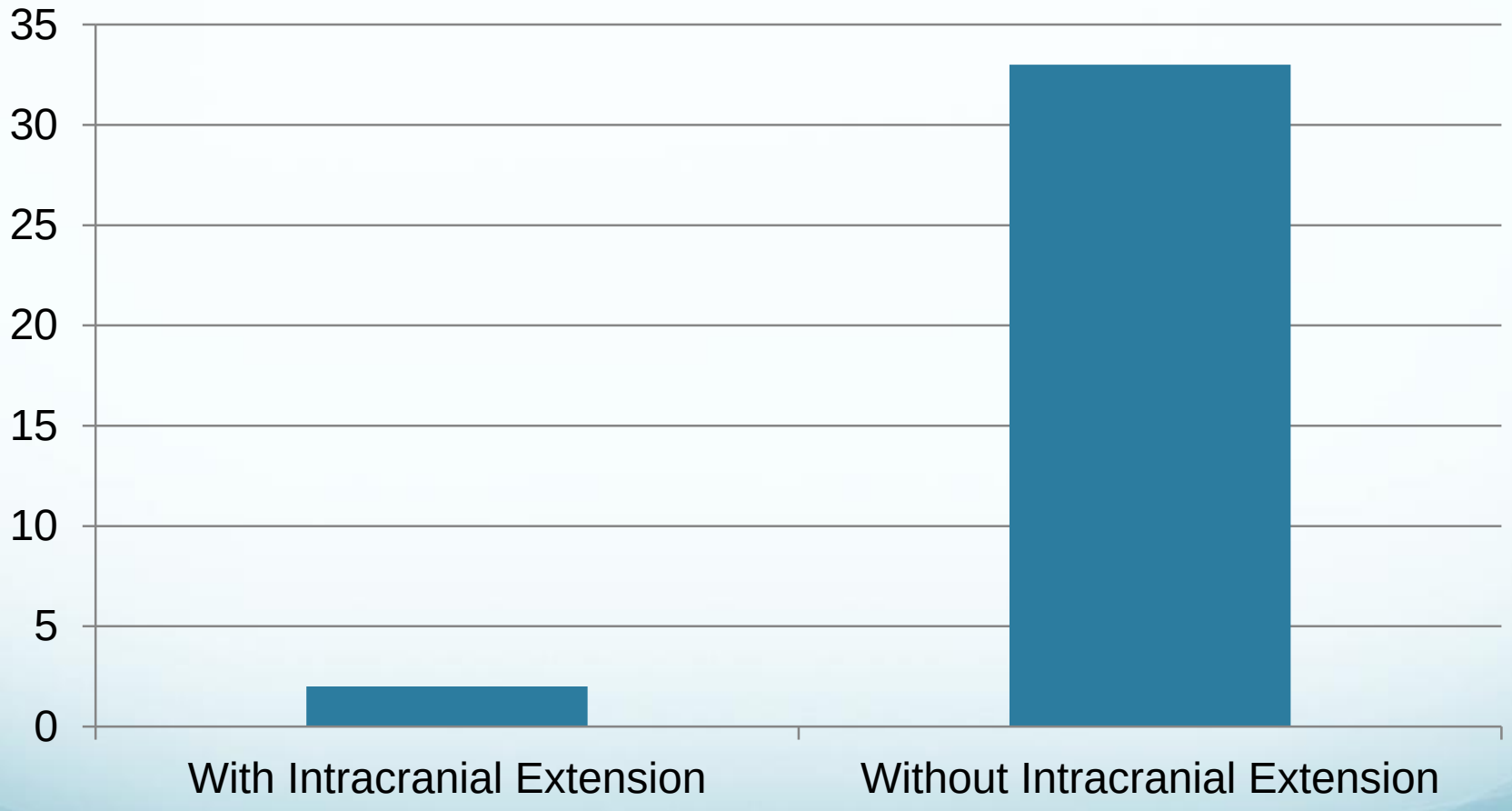
Major Complaints



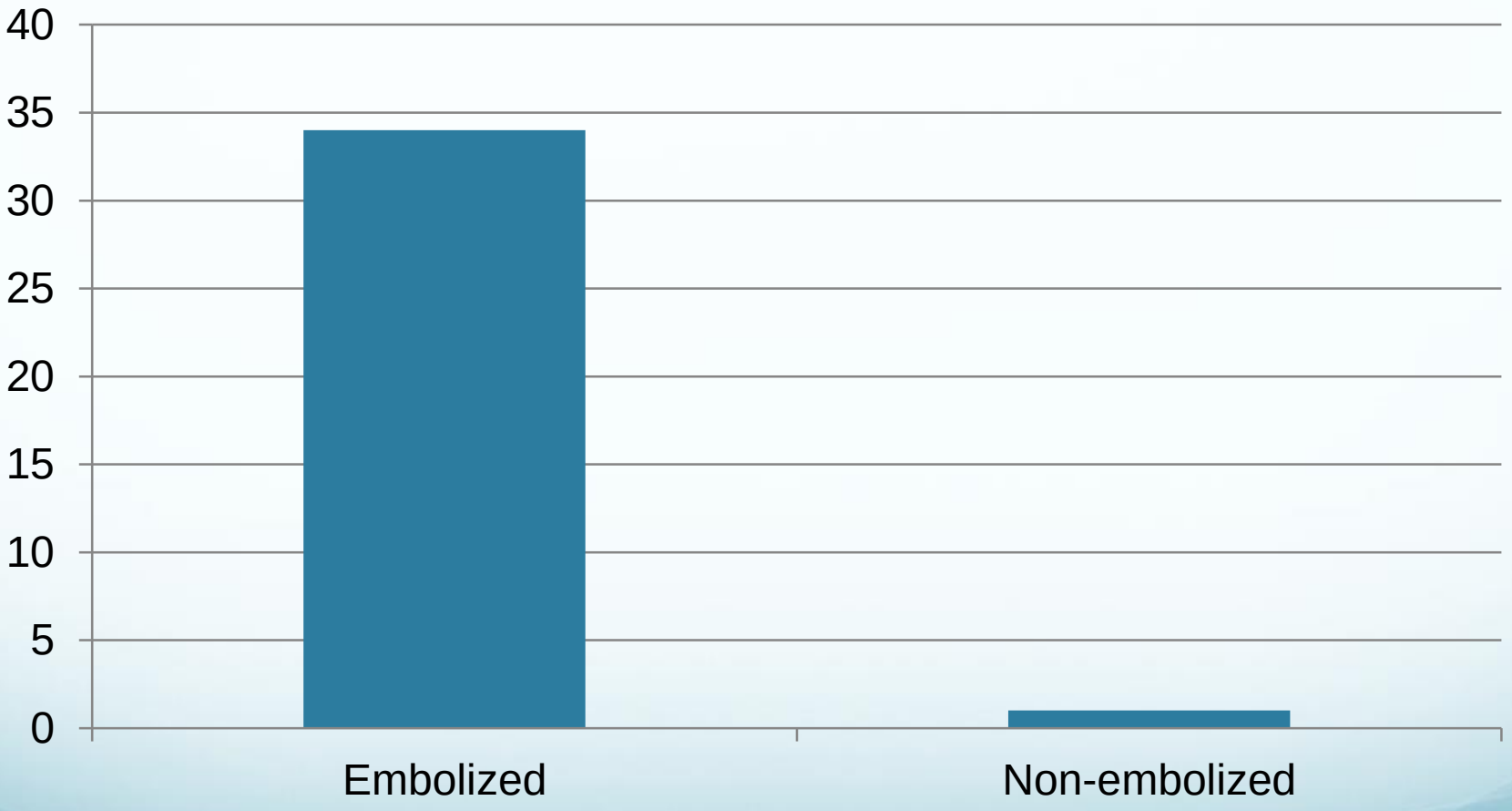
Minor Complaints



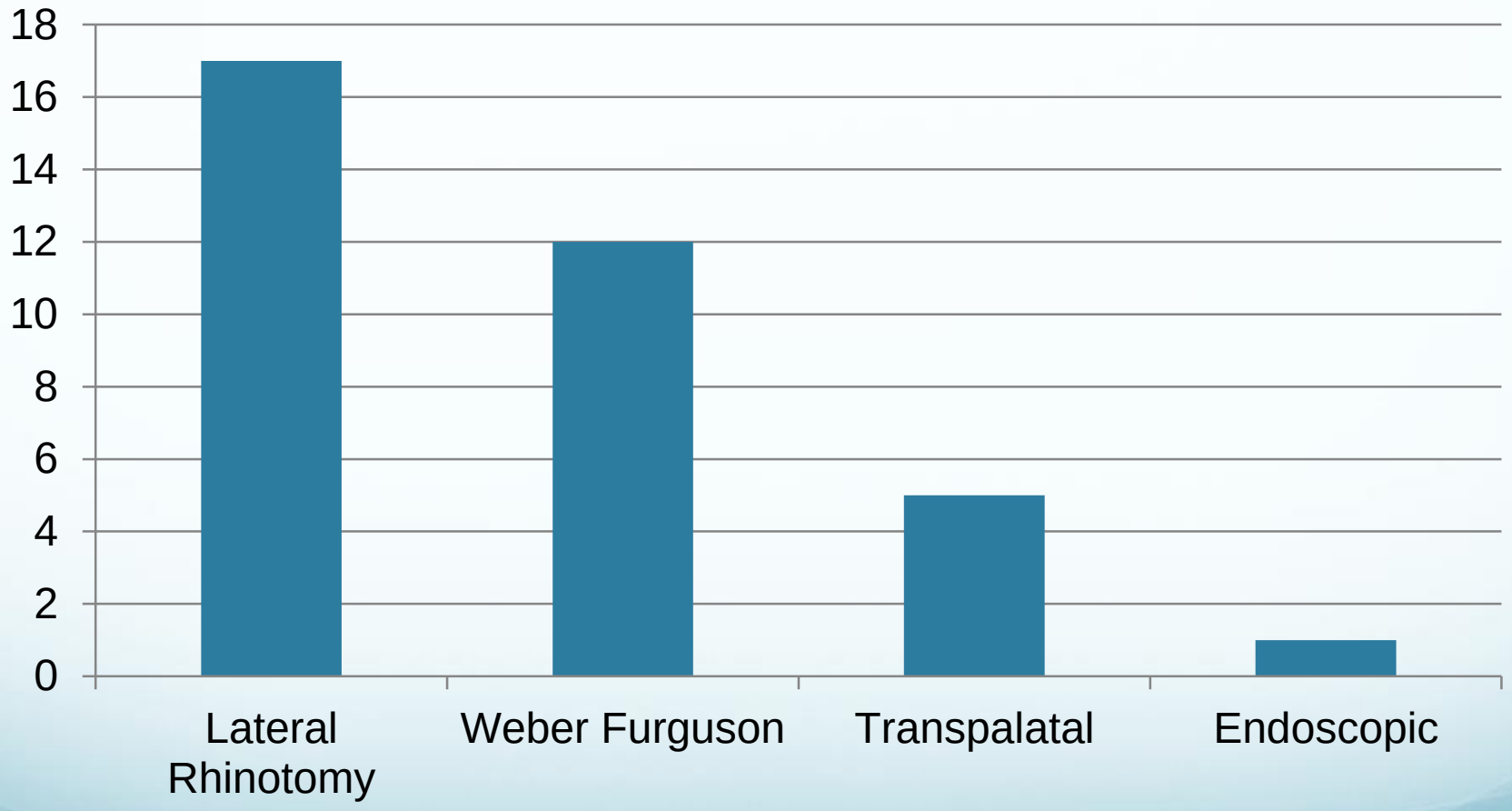
Patients with or without ICE



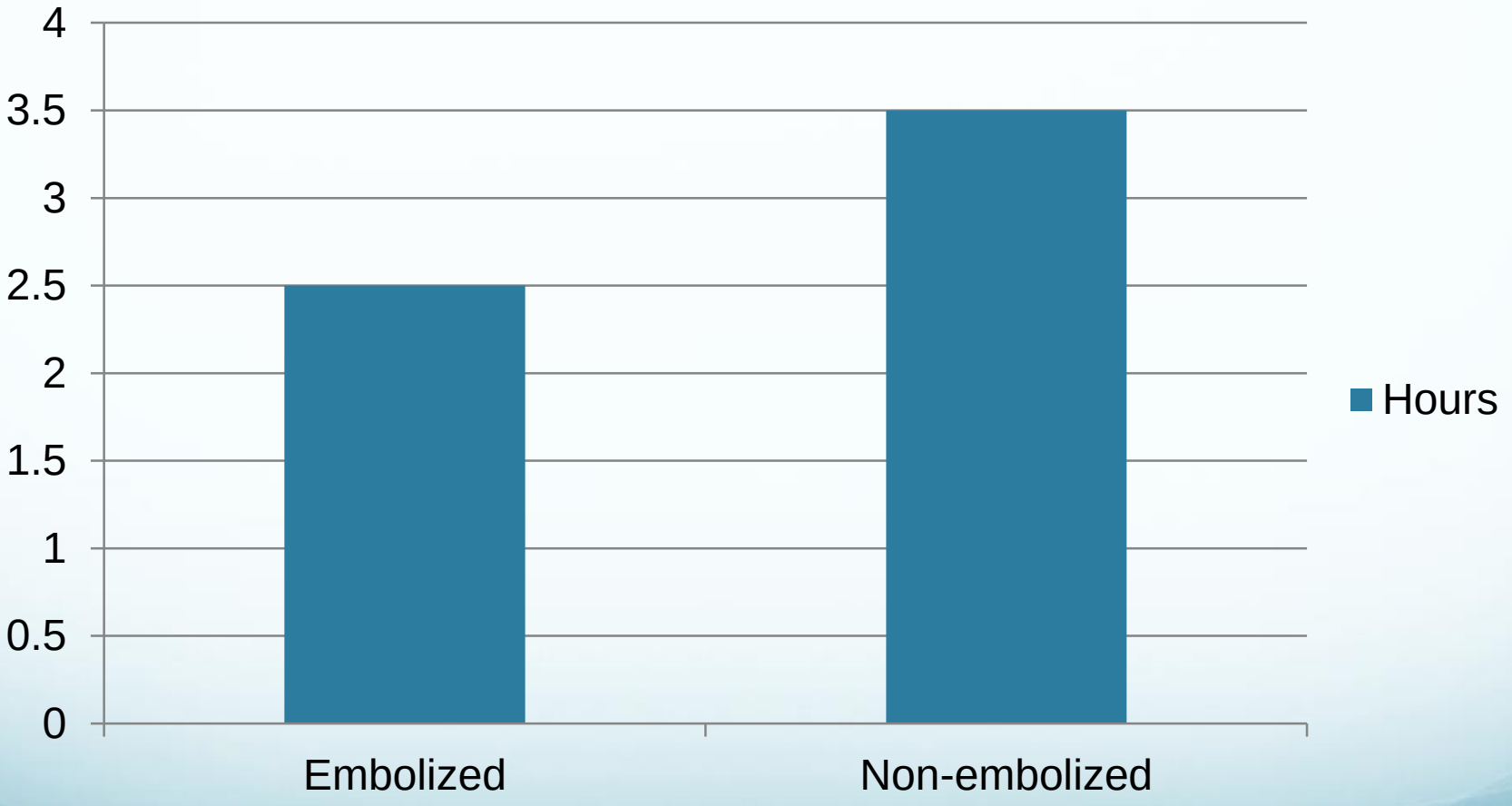
Embolized/Nonembolized Patients



Surgical Approach



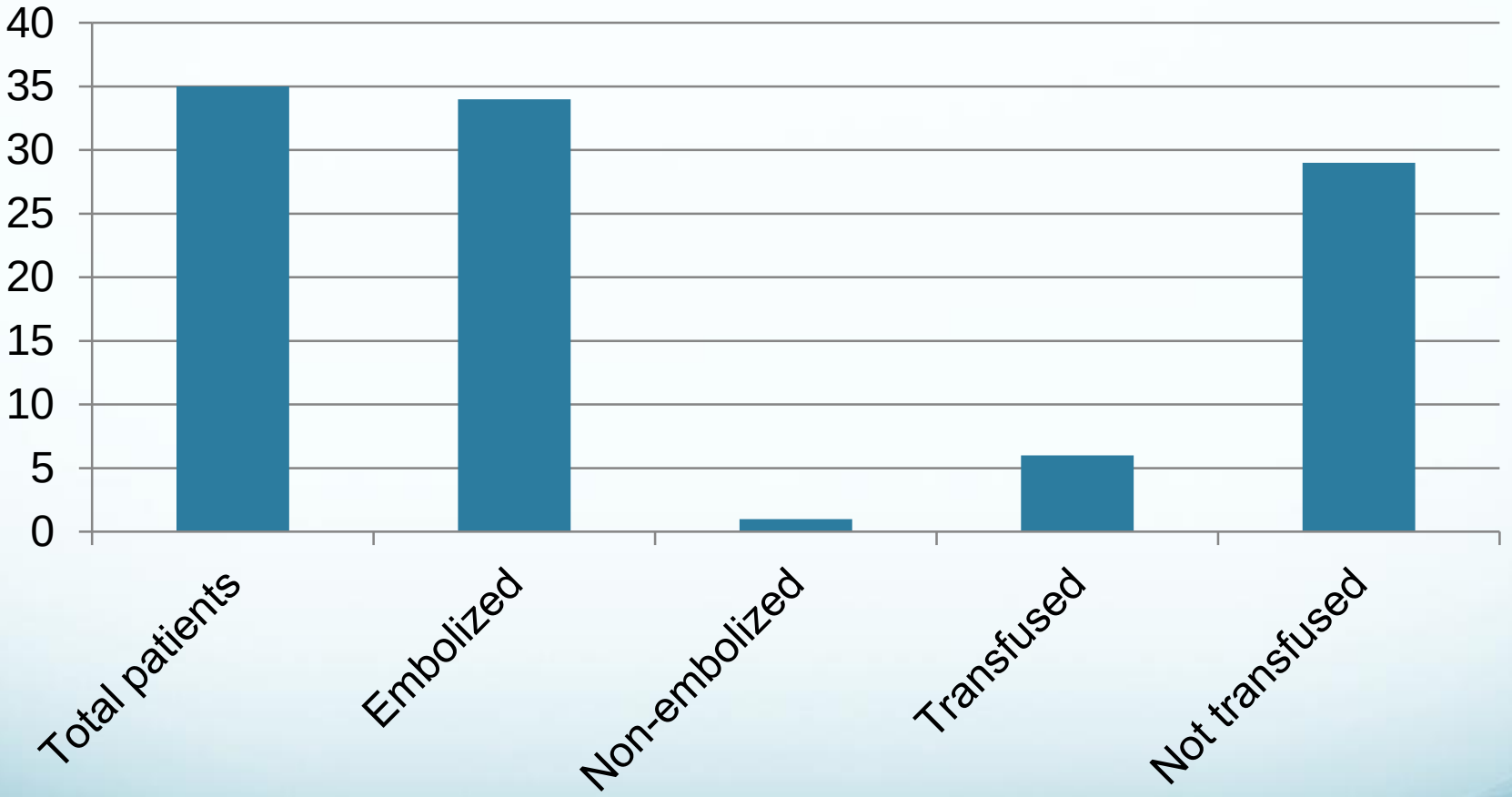
Operative Time



Blood Loss

Blood Loss (mL)	Embolized patients	Non-embolized patients
200-400	11	
400-600	18	
600-700	5	
2000		1

Blood Transfusion



Conclusion

- JNA though rare, yet it is prevalent in our setup in view of the fact that our Hospital is a tertiary care unit.
- Facilities of embolisation are not available in most of the centres in our country
- In developing countries like ours the procedure is cost effective, therefore it is only done at public sector Hospitals
- Surgical resection still remains the Gold standard treatment option

Recommendations

- Repeated follow up serial interval MR imaging should become a standard of care to detect recurrence
- Government should facilitate the procedure in centres other than ours
- Endoscopic treatment is yet to occupy and replace surgical resection in cases of extensive tumours


Thank You!