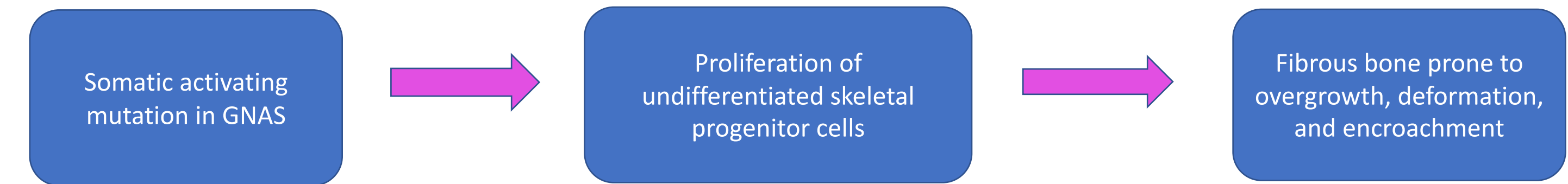


Casting an Eye on Fibrous Dysplasia of the Orbit

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INTRODUCTION

Fibrous dysplasia (FD) is a benign fibro-osseous tumor of growing bone in which fibrous tissue replaces normal medullary bone. [1]



FD is attributed to a somatic mutation in the GNAS protein that results in proliferation of undifferentiated skeletal progenitor cells. The resultant bone is prone to overgrowth, deformation, and encroachment.

Facial asymmetry	Facial pain
Malocclusion	Nasal airway obstruction
Acoustic nerve compression	Optic nerve compression

The symptoms resulting from craniofacial FD are dependent on the amount and location of disease. Amongst the most feared complications is optic nerve compression causing optic neuropathy and threatening visual deterioration and blindness.

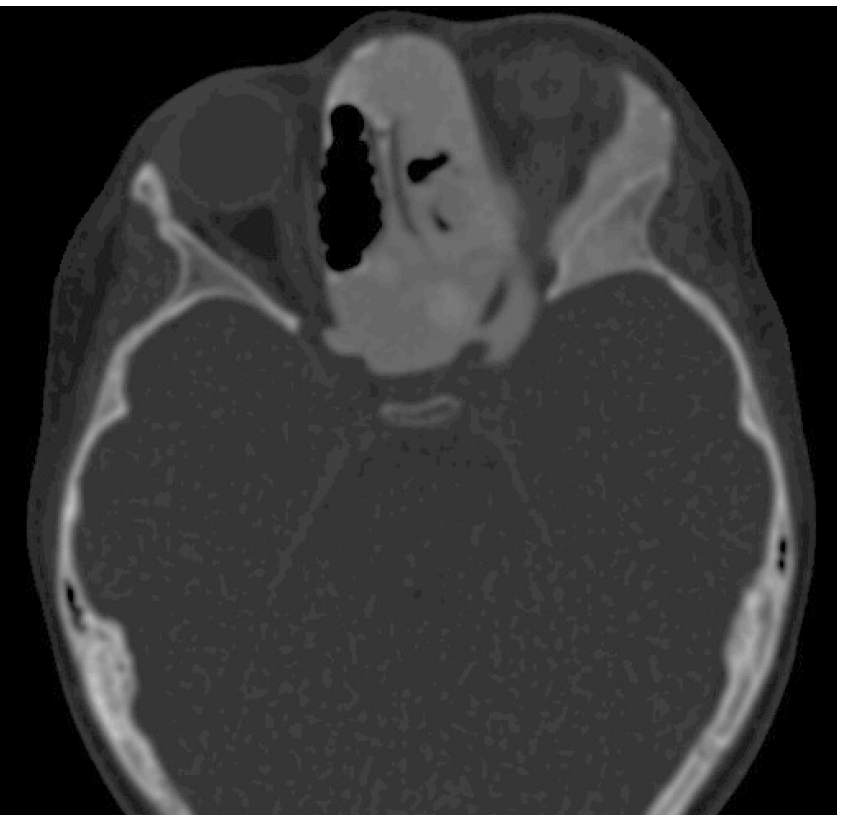


Fig 1. Axial CT image demonstrating left eye FDO

Surgical intervention for FD of the orbit (FDO) (Fig. 1) involves orbital unroofing and optic nerve decompression, yet controversy has surrounded the timing and indication for this procedure (prophylactic vs. therapeutic treatment). [2,3]



Fig 2. CT and clinical image of patient with frontal FDO causing orbital dystopia

OBJECTIVES

The presence of FDO threatens compressive optic neuropathy and irreversible vision loss. The purpose of this study is to retrospectively characterize the natural history of FDO and describe the longitudinal outcomes of 36 patients who underwent stratified surgical management (Fig. 3) at a single institution.

METHODS

Population

- ICD-10-CM codes for monostotic and polyostotic fibrous dysplasia

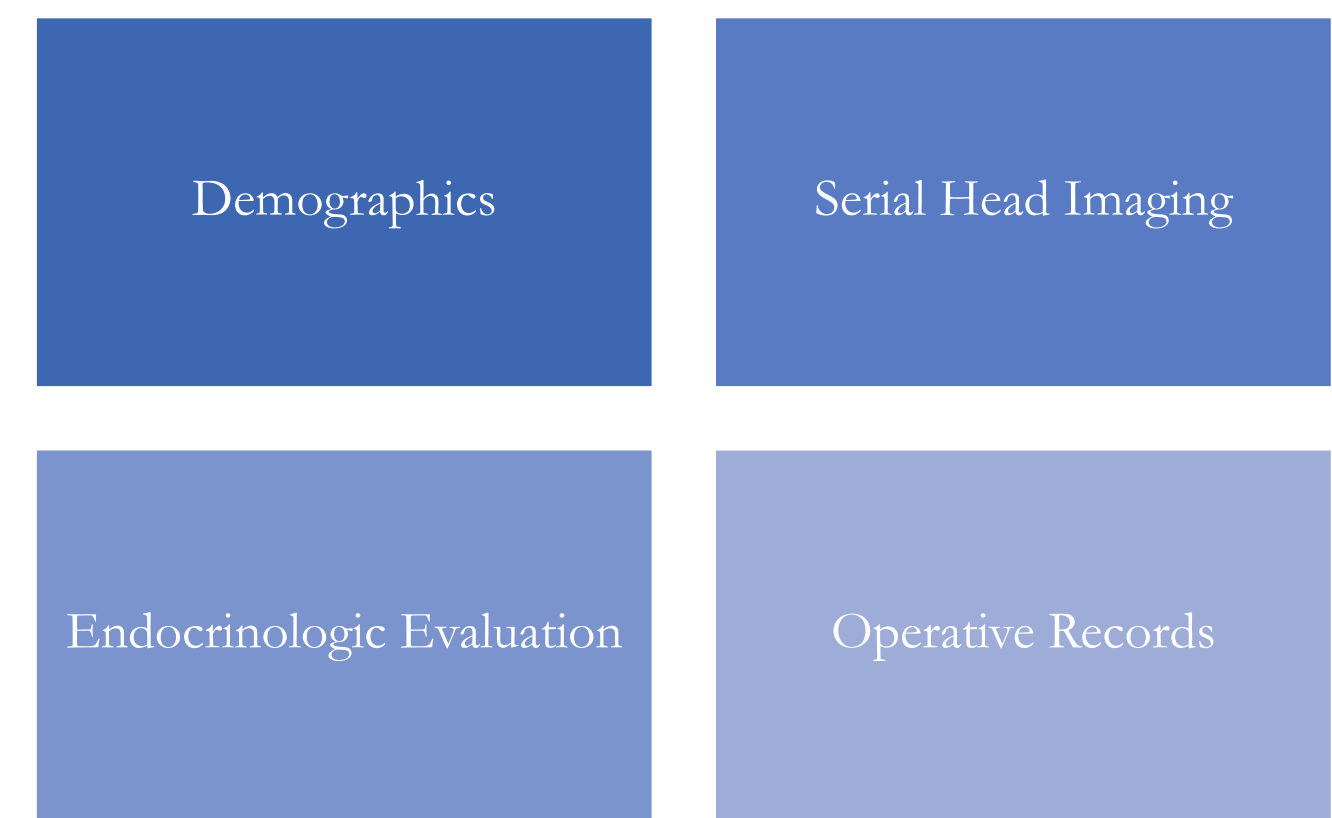
Inclusion criteria:

- Orbital involvement confirmed on CT
- Histopathological confirmation of FD
- Evaluation by neuro-ophthalmology and plastic surgery

Exclusion criteria:

- Non-FD bone tumor
- Lack of orbital involvement
- Insufficient medical records

Variables assessed



Analysis

- Comparative testing**
 - Fisher’s exact test/chi-squared test
 - Independent samples *t*-test
 - Sub-group analysis with McCune Albright Syndrome (MAS) vs. without

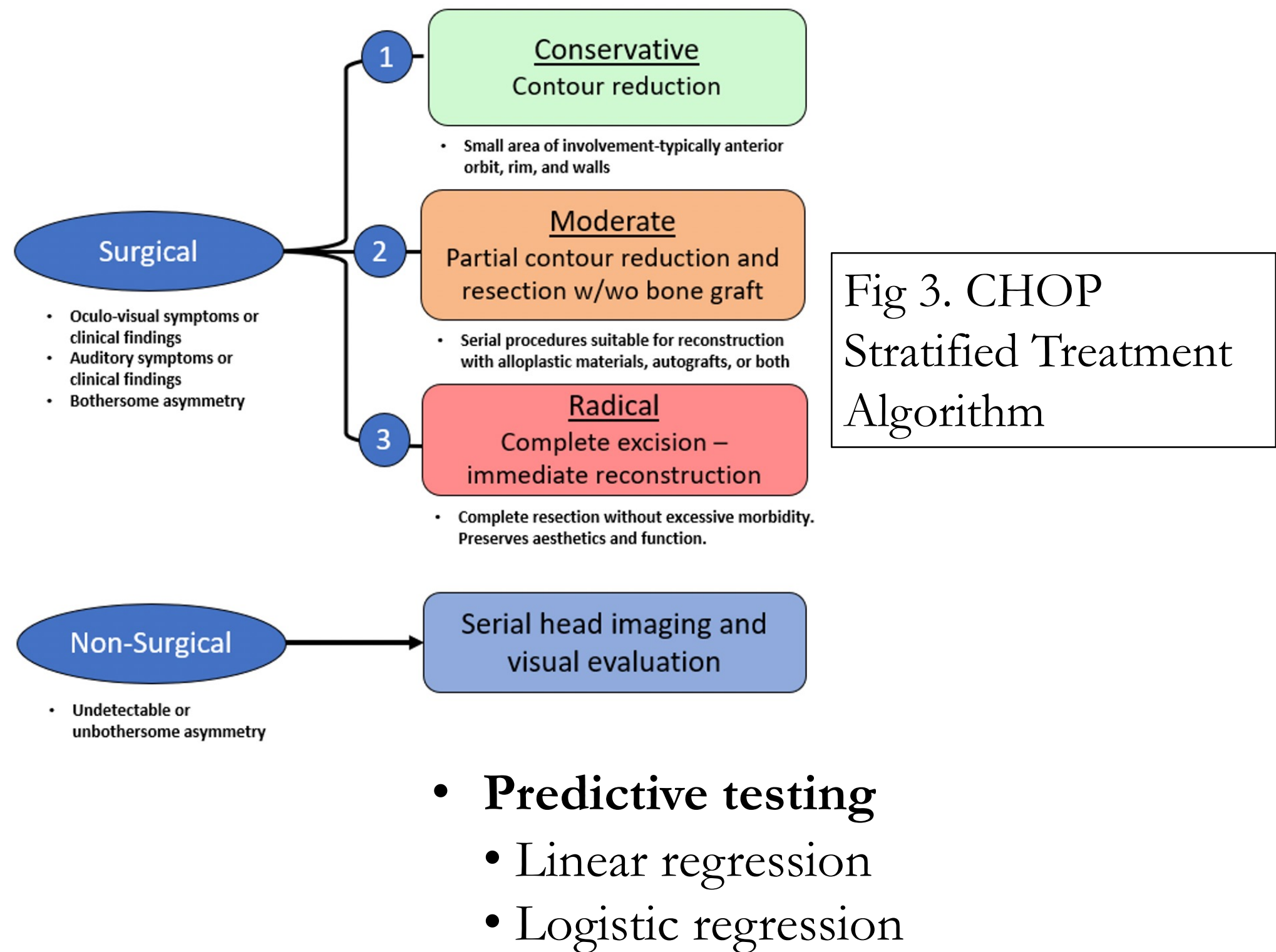
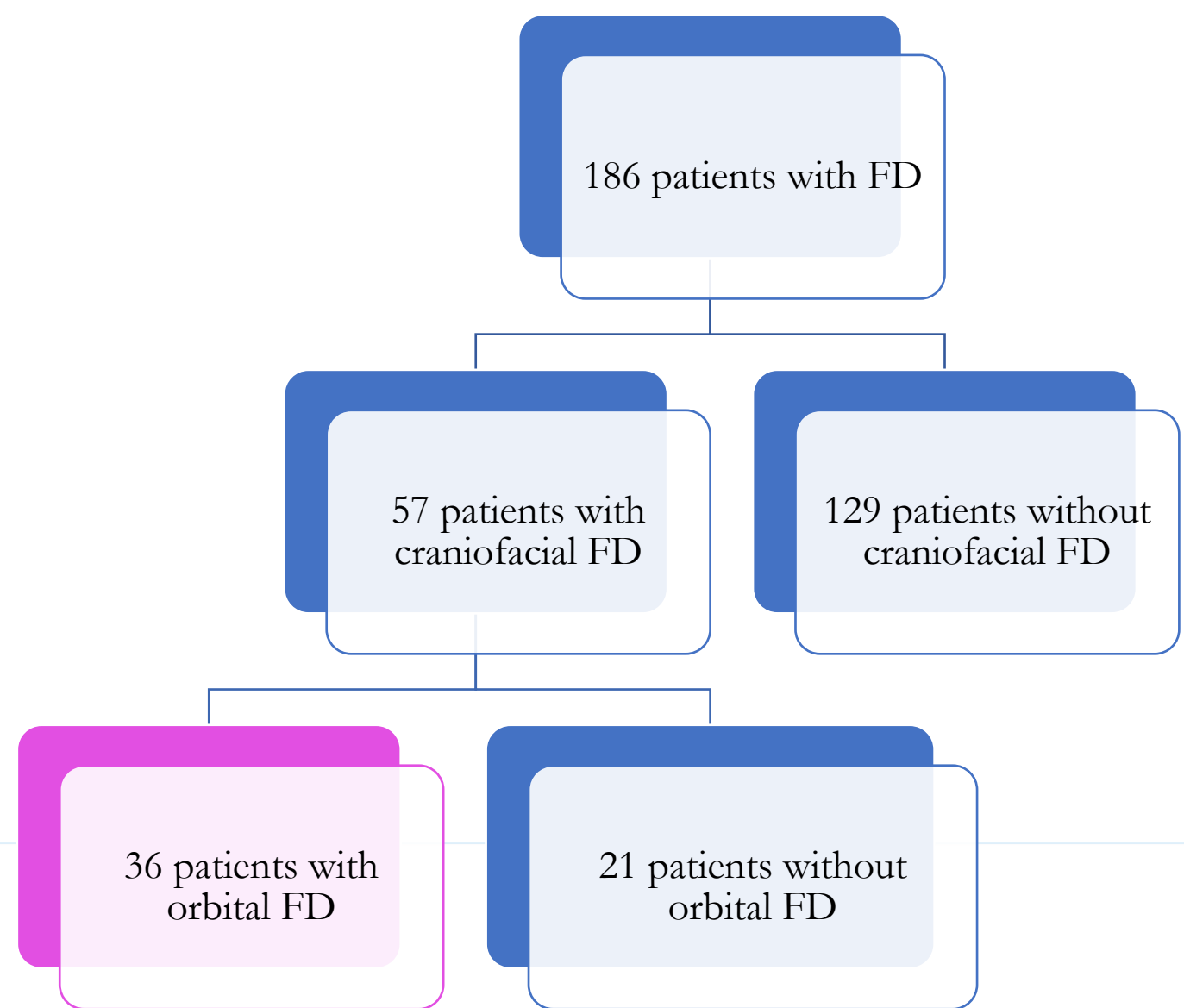


Fig 3. CHOP Stratified Treatment Algorithm

RESULTS



- 164 CT scans analyzed**
- Age at diagnosis**
10.6 ± 4.8 years (0.7 – 18.8 years)
- Timespan covered by CT**
3.8 ± 4.4 years (0.7 – 16.7 years)
- In-person follow-up duration**
5.4 ± 4.7 years (0.15 – 16.6 years)
- Five patients (13.9%) with MAS**

RESULTS CONT.

Imaging of the Orbit

72 orbits belonging to 36 patients

- No facial growth without orbital growth
- 36 orbits (50%) with partially encased optic nerves**
 - 30 optic nerves 100% encased
 - Patients with complete encasement diagnosed significantly younger (8.8 years vs. 12.4, *p* = .025)
- Complete encasement did not predict orbital decompression (*p* = .298)

Surgical Intervention

- 50% of patients**, at least one orbital decompression surgery
- Average age at optic canal decompression: 12.8 ± 3.3 years
- Younger age at diagnosis → **more total operations**
- Younger age at diagnosis → **younger age at optic canal decompression**

McCune Albright Syndrome

- MAS patients diagnosed **significantly younger** (4.1 vs. 11.5), *p* < .001
- No significant difference between groups in...**
 - Number of operations
 - Incidence of optic neuropathy
 - Number of optical canal decompressions

Visual Outcomes

- 100% of patients with stable or improved vision**
- Extraocular symptoms**
 - ptosis/supraorbital nerve palsy (38.5%)
- Younger diagnosis → significantly increased risk of “stable” vision loss**
 - (95% CI [.466 - .960], *p* = .029)
 - MAS was not a predictor in this model

CONCLUSION

No craniofacial FD growth in the absence of orbital FD growth

Progressive disease but complete encasement does not predict optic neuropathy

Therapeutic decompression of the optic canal using our surgical algorithm resulted in favorable visual outcomes

Across the board, younger diagnosis leads to worse outcomes

- ↑ surgeries
- earlier decompression
- worse visual outcomes

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