

Glaucoma in children with facial port wine stain

By:

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Disclosure:

The authors have no financial interest in any of the material stated in the study.

Introduction:

- Facial port-wine stain (PWS) is a manifestation of **Sturge-Weber Syndrome (SWS)**, one of the **neurocutaneous syndromes**.
- Glaucoma is the most common ophthalmic association with SWS, incidence **30% -70%**.
- **Mechanism:**
 1. Goniodysgenesis.
 2. Elevated episcleral venous pressure.
 3. Premature aging of the trabecular meshwork-Schlemm's canal complex.
 4. Aqueous hypersecretion by the ciliary processes.

Aim of study:

- Report the **incidence** and **clinical presentation** of glaucoma in children presenting with facial PWS.
- Report the **rate of conversion to glaucoma** among children with facial PWS initially diagnosed as not glaucomatous or suspects.
- Report the **results of the surgical treatment** of glaucoma associated with facial PWS.

Subjects and methods:

- Type: Retrospective chart review.
- Children with facial PWS referred to the pediatric ophthalmology practice of the Ophthalmology Department, Alexandria Main University Hospital, from **December 2005 to December 2016**.
- Records reviewed for:
 1. Patient demographics.
 2. Results of clinical ophthalmic examination and systemic examination findings on first presentation of the child.
 3. Results of examination at the final follow up visit.

Subjects and methods:

- Clinical stratification of study children eyes was done on initial presentation and at the final visit into 'glaucoma', 'glaucoma suspects' and 'no glaucoma'.
- **GLAUCOMA** = 2 or more criteria of an IOP above 15 mmHg under GA and/or a cup/disc ratio above 0.3 and/or corneal pathology diagnostic of elevated IOP (oedema, Haab's stria, broad limbus, enlarged diameter).
- **GLAUCOMA SUSPECT** = Only one of these features.
- **NO GLAUCOMA** = Absence of all these features.
- Most importantly, **progression over time** = "glaucoma", and a **stationery condition** = "glaucoma suspect" or "no glaucoma".

Subjects and methods:

- All glaucomatous eyes were operated by combined trabeculotomy-trabeculectomy ith mitomycin C and a prophylactic inferior sclerotomy was performed at the time of surgery.
- **Success** of surgery was defined as an **IOP below 15 mmHg with absence of any vision threatening complications.**
- Qualified success : successful IOP control with the need of IOP-lowering therapy.
- Complete success : successful IOP control without the need of IOP-lowering therapy.

Results: (44 eyes of 22 children)

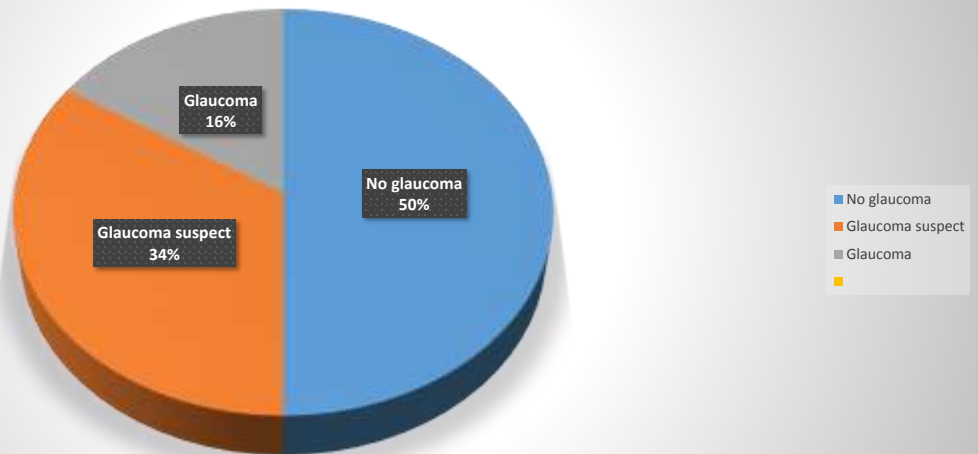
Demographic criteria	
1. Gender	
• Males	10 (45%)
• Females	12 (55%)
2. Age at presentation (mean±SD, range, Median) in months	18.2±33.9, 1.8 – 120.5, 4.0
3. Parental consanguinity	6 (27%)
4. Term at delivery	
• Full term	21 (95%)
• preterm	1 (5%)
5. NICU admission	7 (32%)

Results:

Clinical criteria	
1. Port wine stain	
• One division of CN V	7 (32%)
• More than one division	15 (68%)
• Involving trunk and extremities	5 (23%)
2. Leptomeningeal angioma	1 (5%)
3. Convulsions	5 (23%)
4. Hazy cornea at presentation	2 (9%)
Follow up (mean±SD, range, Median) in months	16.1±24.8, 0.0 – 89.8, 3.1

Results:

Clinical characteristics at initial visit



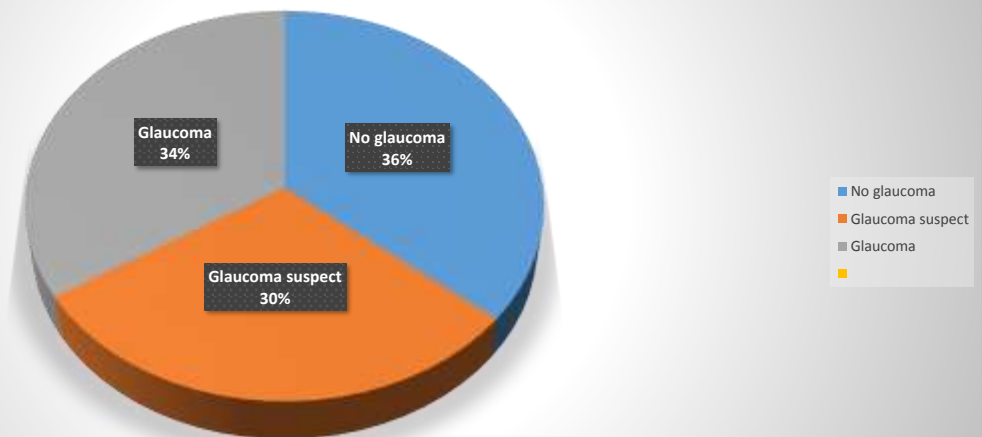
Results:

Clinical characteristics at initial visit

	Glaucoma eyes (7 (16%))	Glaucoma suspects (15 (34%))	No glaucoma (22 (50%))
IOP (mean±SD, range, Median) (mmHg)	16±6.2, 6 – 30, 16	11.4±7.3, 4 – 30, 11.0	8.2±2.6, 4 – 12, 8.0
Corneal Diameter (mean±SD, range, Median) (mm)	12.0±0.7, 10.5 – 13, 12	11.4±0.9, 10.0 – 13.0, 11.5	11.1±0.4, 10.5 – 12, 11
Cup/disc ratio (mean±SD, range, Median)	0.5±0.3, 0.1 – 0.9, 0.5	0.3±0.3, 0.0 – 0.9, 0.3	0.1±0.1, 0.0 – 0.5, 0.1
Axial length (mean±SD, range, Median) (mm)	21.4±1.6, 19.3 – 25.3, 21.0	21.0±2.9, 17.9 – 25.3, 21.0	20.6±1.6, 18.4 – 23.1, 20.7

Results:

Clinical characteristics at final visit



Results:

Clinical characteristics at final visit

	Glaucoma eyes (15 (34%))	Glaucoma suspects (13(30%))	No glaucoma (16 (36%))
IOP (mean±SD, range, Median) (mmHg)	20.6±5.1, 15 – 32, 18.0	13.6±5.4, 2 – 24, 12.0	7.5±1.7, 4 – 10, 8.0
Corneal Diameter (mean±SD, range, Median) (mm)	13.0±0.9, 11.0 – 14.0, 13.0	12.0±0.8, 10.5 – 13.0, 12.0	11.3±0.6, 10 – 12, 11.0
Cup/disc ratio (mean±SD, range, Median)	0.7±0.2, 0.4 – 1.0, 0.8	0.5±0.3, 0.0 – 0.9, 0.4	0.1±0.1, 0.0 – 0.3, 0.1
Axial length (mean±SD, range, Median) (mm)	23.3±1.8, 19.1 – 25.7, 23.9	22.0±1.7, 19.1 – 24.8, 22.5	21.0±1.0, 18.7 – 22.7, 21.1

Results:

Clinical stratification of study eyes

Eyes initial visit no glaucoma converted to glaucoma in last visit (n,%)	1 (2.3)
Eyes initial visit no glaucoma converted to glaucoma suspect in last visit (n,%)	6 (13.6)
Eyes initial visit no glaucoma remained no glaucoma in last visit (n,%)	15 (34)
Eyes initial visit glaucoma suspect converted to glaucoma in last visit (n,%)	7 (16)
Eyes initial visit glaucoma suspect converted to no glaucoma in last visit (n,%)	1 (2.3)
Eyes initial visit glaucoma suspect remained glaucoma suspect in last visit (n,%)	7 (16)
Eyes initial visit glaucoma confirmed glaucoma in last visit (n,%)	7 (16)

Results:

Clinical Examination Data of operated eyes (11)

Mean (SD, range, median)	Preop.	1m	3m	6m	9m	12m
IOP (mmHg)	18.2 (4.8, 12 – 26, 16)	8.0* (2.4, 6 – 12, 8)	6.3* (4.9, 3 – 12, 4)	5.0 (3.6, 1 – 8, 6)	8.0* (4.0, 4 – 12, 8)	9.0* (8.3, 4 – 32, 6)
Corneal Diameter (mm)	12.9 (0.9, 11 – 14, 13)	12.4 (0.4, 12 – 13, 12.5)	11.8 (0.3, 11.5 – 12, 12)	12.5 (0.0, 12.5 – 12.5, 12.5)	12.7 (0.3, 12.5 – 13.0, 12.5)	12.7 (0.4, 12 – 13.5, 12.5)
C/D ratio	0.7 (0.2, 0.1 – 0.9, 0.8)	0.3 (0.3, 0 – 0.8, 0.3)	0.4 (0.3, 0 – 0.6, 0.6)	0.3 (0.3, 0 – 0.6, 0.2)	0.3 (0.4, 0 – 0.7, 0.3)	0.4 (0.3, 0 – 0.9, 0.3)
AL (mm)	22.6 (1.7, 19.5 – 24.2, 23.3)	22.6 (1.5, 20.7 – 23.7, 23.6)	21.6 (1.6, 19.2 – 23.5, 21.4)	21.1 (1.4, 19.7 – 22.4, 21.2)	23.6 (0.5, 23.1 – 24.1, 23.5)	22.8 (1.9, 19.7 – 25.2, 23.1)

Results:

- The success rate in the operated eyes was around **90%**.
- IOP reduction in around **90%** and reversal of optic nerve cupping in **73%** of operated eyes.
- **No intraoperative complications** occurred in any of the operated eyes.
- Two eyes with a choroidal haemangioma developed an **exudative choroidal detachment** postoperatively.

Conclusion:

1. **Glaucoma** is a significant ocular hazard in children with facial PWS.
2. Glaucoma may not be evident on initial presentation of the child with facial PWS and **may present late (rate of conversion 28%)**.
3. The presentation is usually with a **clear cornea**.
4. Surgical intervention is associated with a **high success rate and a low rate of complications**.

