Management of childhood glaucoma associated with Sturge-Weber syndrome

Greslehner R, Helbig H, Oberacher-Velten I
Department of Ophthalmology, University Medical Center Regensburg, Germany

BACKGROUND:
- rare, congenital, sporadic disorder
- incidence 1: 50 000
- **Vascular malformations:**
  - Skin
    - port-wine stain
  - Central nervous system
    - seizures, transient focal deficits, mental retardation
  - Eye
    - hemangiomas of the conjunctiva, episclera, iris and choroid
- **glaucoma in 30 - 70%**
- **high risk** of severe complications
- intraoperative **expulsive hemorrhage**
- **massive choroidal effusion**
- **serous retinal detachment.**

RESULTS:

Clinical findings:
- facial port-wine stains:
  - 2 patients: V1 + upper eyelid,
  - 3 patients: V1, V2, V3 + both eyelids
- buphthalmus: all
- dilated episcleral vessels: all
- diffuse choroidal hemangiomas: 4 eyes.
- irisheterochromia: 3 eyes

Treatment:
- Glaucoma surgery was performed in four children.
- One child was treated with antiglaucomatous medication alone.

Complications:
- **Reversible uveal effusion** and subluxation of the lens appeared postoperatively in one eye,
- **persistent serous retinal detachment** occurred 3 years after surgery in another eye (with an intraocular pressure of 10 mmHg).

Both complications were found in eyes with diffuse choroidal hemangioma.

METHODS:
- retrospective case series
- **5 children/ 5 eyes**
- mean age **5.6 years**
- secondary glaucoma associated with Sturge-Weber syndrome.

Ocular findings, treatment modalities, intraocular pressure and complications were assessed.

CONCLUSION:
Management of glaucoma associated with Sturge-Weber syndrome is difficult and controversial. Medical treatment often does not decrease intraocular pressure sufficiently. When planning surgical intervention an increased risk of severe complications has to be considered, especially in the presence of diffuse choroidal hemangioma.

REFERENCES:

No financial interests
Contact: greslehner@eye-regensburg.de