ters), which would smooth out the lows as well as the highs. Having one year of endowment money, already in place as a cushion, is also key in years such as this one. It is crucial to have reserve funds available for managing deficits across the department.

Randall Olson talked about clinical income. It is apparent that the number of cosmetic procedures, such as refractive and cosmetic plastics, is steadily decreasing. The number of refractive surgeries could dip by as much as 50% during this down cycle. Elective procedures are generally being performed at the same rate in most parts of the country but the amount of self-pay is likely to increase as patients lose their insurance. Managing the self-pay category so that some of that revenue is captured will be important so that overall collections on our activity do not drop drastically.

Randall Olson also spoke about maintaining a development effort in spite of the times. This is not a time to decrease our effort in any way. Many will give future commitments rather than donating right away. It is important to let them know that just because they are in financial difficulty does not mean cultivation or stewardship will change. It is also important to remember that not everybody is in a down cycle. Some people are actually in a better position to help right now and require continued cultivation and stewardship. Also, those who cannot give at the same level as they have in the past may be willing to give a decreased amount. Every little bit helps.

Peter McDonnell talked about maintaining a research effort. Unfortunately, we are in the midst of a perfect storm. Flexible funding has decreased institutionally, fund endowments have imploded, fundraising is increasingly difficult, and NIH funding rates are poor. Now is the time to look for every possible funding opportunity, both inside and outside the institution (DOD, Pharma, etc). Tough decisions about faculty who have remained unfunded must be made. While flexible funds are provided to those who are worth saving, without the necessary reserves, the department may not be able to continue maintaining a viable research effort. In this case, decisions about overall cuts that come from a committee of researchers will be much easier to enact than those dictated by the Chair alone.

Stickler Involved People

BY JAN HELFER

STICKLER SYNDROME (a "zebra" syndrome affecting the eyes):

- is a genetic, autosomal dominant disorder caused by a collagen gene mutation
- is the leading cause of retinal detachment in children and the most common cause of inherited rhegmatogenous retinal detachment
- may possibly be the most common connective tissue disorder in North America and Europe but is frequently undiagnosed or misdiagnosed
- is a progressive disorder with incomplete penetrance, varied expression, and severity
- affects not only the eyes, but also causes auditory, skeletal and/or craniofacial abnormalities
- is often confused with other syndromes such as Wagner, Marshall, or Marfan
- does not affect lifespan or mental capacity

Dr. Gunnar B. Stickler (1965) originally termed the syndrome "hereditary progressive arthro-ophthalmopathy" because he associated severe sight deterioration with joint changes in several generations of one family.

Three gene mutation locations are currently known—Stickler Type I (COL2A1), Stickler Type II (COL11A1) and Stickler Type III (COL11A2). Types I and II have distinctive ocular characteristics, with Type III affecting joints and hearing, but not vision. In addition, there is at least one other type of Stickler syndrome with an unidentified gene mutation location.

The prevalence of Stickler syndrome is 1 to 3 per 10,000. Early identification is critical, especially due to the high instance of retinal detachment in children (20% before age 10 as reported in Stickler, Hughes, Houchin, 2001) and the inability of children to identify and verbalize symptoms. Detachments may, therefore, be inoperable by the time they present to the physician.

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STICKLER SYNDROME

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Stickler syndrome may be diagnosed by either genetic testing or diagnostic criteria. The former is complicated by the genetic heterogeneity of the mutation. The diagnostic criteria, published by the National Institutes of Health in 2005, have been found to be 98% reliable for Type I Stickler syndrome (75% of all Stickler patients). The criteria evaluate characteristic ocular, auditory and skeletal abnormalities, molecular data, and family history. Common ocular features of Stickler syndrome include:

- high myopia (–5 to –18 diopters) often present at birth
- spontaneous retinal detachment, usually a giant tear
- congenital abnormality of the vitreous
- pre-senile cataracts (wedge or fleck)
- open-angle glaucoma (less common)
- chorioretinal degeneration—lattice formation, holes, tears
- strabismus/astigmatism

Patients with Stickler syndrome are likely to have the following needs related to eye care:

- corrective lenses
- frequent (at least annual) eye exams, including "evaluation under anesthesia" for infants and young children
- prophylactic laser or cryo treatment (see below)
- regular eye pressure readings and education of the patient as to normal range
- avoidance of contact sports

Dr. Martin P. Snead's *Ophthalmology* article (Jan. 2008) outlined the long-term benefits of prophylactic surgery and recommendations for treatment technique. Snead recommends placement of the therapeutic lesions to be just posterior to the ora serrata. Lesions placed equatorially or too posterior are more likely to fail, thereby allowing a giant tear to progress to detachment. Prophylactic treatment is recommended early as Stickler infants have been known to experience a giant tear by 18 months of age.

Suspicion or diagnosis of Stickler syndrome should lead the physician to not only provide the appropriate treatments, but also to make a referral to a geneticist. Further patient information is available from Stickler Involved People (SIP), a not-for-profit volunteer organization with a mission to support those affected by the syndrome. SIP strives to raise the awareness of health care providers and increase their knowledge about this disorder. To that end, SIP sponsors an annual three-day conference with physician speakers who discuss the latest knowledge and treatment of the syndrome.

For more information, including a brochure, presentation, and 30-minute DVD, please visit www.sticklers.org. For a hard copy of these materials, e-mail sipbrochure@yahoo.com with your mailing address. Additional questions about the syndrome or SIP services can be directed to sip@sticklers.org or to jdhelfer@yahoo.com.