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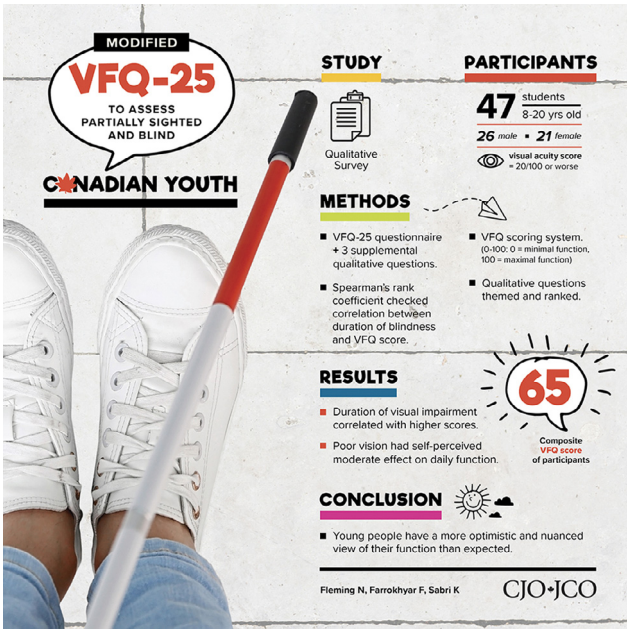

Assessment of the visual function of partially sighted and blind Canadian youth using the VFQ-25 questionnaire: a resident's perspective

Gareth Mercer, PGY-3

Patient Reported Outcome Measures (PROMs) are tools (typically questionnaires) that measure aspects of illness, functional status or therapeutic effects as reported directly by patients.[1] With the movement to make health care more patient centered, PROMs are gaining an increasingly important place in clinical practice and medical research.[1] In Ophthalmology, PROMs tend to address one or more of the following domains: vision related quality of life, functional vision, or severity of symptoms like ocular pain.[1,2]

Many of the conditions leading to childhood visual impairment in high-income countries are not preventable or reversible. In this setting, PROMs can be used to guide rehabilitation efforts to mitigate the effects of visual impairment on social and occupational functioning.[2] It is accepted that children as young as 7 years old can validly and reliably respond to appropriately-designed PROMs.[2] However, it is essential that such PROMs use developmentally appropriate language and assess activities and experiences that are relevant to children.[2] These added methodological challenges mean that few validated PROMs are currently available for use with visually impaired children.[3]

In the current issue, Fleming and colleagues evaluate a PROM called the Visual Function Questionnaire (VFQ) among a sample of 47 visually impaired Canadian youth between 8 and 20 years of age.[4] The VFQ is the most commonly used vision-related PROM in the world.[1] It was developed from focus group discussions involving adults with a variety of eye conditions and is intended to measure the quality of life impacts of "all cause" visual impairment.[5] The shortened version of the VFQ includes 25 questions covering: general health, general vision, near and distance vision, peripheral vision, colour vision, driving, role limitations, dependency, social function, and mental health.[5]

Despite having advanced vision loss (most with acuities of 20/200 or worse), the children and youth in Fleming and colleagues' sample reported relatively high VFQ-25 scores compared to cohorts of adults with similar levels of acuity. In particular, they reported comparatively high scores in the social functioning and mental health domains. Interestingly, children with congenital or early onset visual impairment reported higher levels of vision related quality of life than those with more recent onset visual impairment, possibly signifying better adaptation or greater acceptance of their levels of visual functioning. A limitation of these findings is that the study did not specifically address whether the VFQ-25 was developmentally appropriate for the age group studied, nor whether the respondents found the items on the questionnaire to be generally relevant to their lives.

While further work is necessary to validate the VFQ for use with children, clinicians and researchers wishing to include these types of measures in their work may find helpful information in Tadić and colleagues' systematic review of PROMs for paediatric ophthalmology[3] and in the Patient-Reported Outcome and Quality of life Instruments Database (https://eprovide.mapi-trust.org/about/about-proqolid).

Referenced article: https://www.canadianjournalofophthalmology.ca/article/S0008-4182(19)30036-5/fulltext

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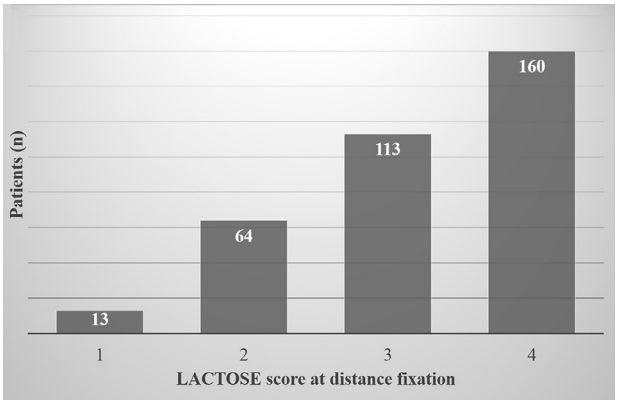

Control is key: Predicting surgical success in intermittent exotropia

Laura Donaldson, PGY-5

Intermittent exotropia is a common form of childhood strabismus that is usually seen in association with good visual acuity and initial development of binocular vision. Surgical treatment is common, though the precise indications for and timing of intervention are variable. One reason for this is that outcomes are often unpredictable. Efforts to correlate pre-operative factors such as age, angle of deviation, refractive error, fusional ranges, or stereoacuity with surgical success rates have yielded disappointing or conflicting results.[1,2] In this issue, Moon and colleagues describe a simple test for predicting surgical outcomes in intermittent exotropia.[3]

The authors describe a large cohort of 350 South Korean patients with basic type intermittent exotropia undergoing bilateral lateral rectus recession by a single surgeon for an angle of at least 20 prism diopters (PD). They describe a novel scoring system based on degree of control, which is abbreviated as LACTOSE: Look And Cover, then Ten seconds of Observation Scale for Exotropia. A maximum score of 4 was assigned if exotropia was manifest in 10 seconds of fixation. If not, an alternate cover test was performed for 10 seconds before the patient was asked to refixate, then a score from 0 (exophoria) to 3 (exotropia manifest for the entire 10 seconds post-alternate cover testing) was assigned. Two scores were done three months apart by a single surgeon, who was masked to their previous score.

Surgical success was defined as an alignment between 10 PD of exodeviation and 5 PD of esodeviation at both distance and near at one year. In the success group, average LACTOSE scores were 3.11 ± 0.89 at distance and 1.94 ± 1.11 at near, compared with 3.54 ± 0.65 and 2.38 ± 1.11 respectively in the failure group. This difference was statistically significant between the two groups while age, angle of deviation, stereoacuity, and refraction were not.

This work describes a new tool for strabismus surgeons that can be used when attempting to answer a question that is constantly asked by parents: will surgery be effective for my child? It is quick, easy to perform in clinic, and shows promise in discriminating between the two categories of surgical success versus failure. It does not appear to capture all of the elements that predict success, as many patients with a maximum score of 4 went on to have good post-operative outcomes. It would be interesting to know whether LACTOSE scores also correlate with outcomes in a natural history study of untreated exotropia.

Though an impressive cohort, only 169 patients (48%) completed a follow-up assessment at 1 year and were included in the analysis of surgical success. Decompensation often occurs many years post-operatively, and so reassessment of the study patients even further out would be informative. The overall one-year success rate reported was excellent (78%), however, was defined on pure motor parameters. Considering sensory outcomes as well as motor further characterizes functional vision and results in a much smaller proportion of patients being classified as a good surgical result,[4] potentially altering study conclusions substantially.

Surgeons should consider evaluating and documenting degree of control in patients with intermittent exotropia preoperatively, and the authors provide an efficient means to do so. This work adds to the understanding of this common but poorly understood form of strabismus and opens up exciting possibilities for future research in predicting outcomes and influencing surgical planning.

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Papilledema in the pediatric ambulatory clinic

Matthew D. Benson, PGY-4

Papilledema is a term reserved for optic nerve edema secondary to raised intracranial pressure (ICP). In children, elevated ICP is considered when a lumbar puncture opening pressure is >28cm H_2O.[1] While a diagnosis of true papilledema is rare in pediatric patients referred to an ambulatory ophthalmology clinic with a suspicion of papilledema,[2] a careful fundus examination is warranted to avoid missing a potentially life-threatening disease process.

In this issue, Hyde et al. retrospectively examined the etiologies of confirmed cases of papilledema in a pediatric population from 1996 to 2018. Their study population included primarily ambulatory outpatients from two separate sites. Interestingly, the literature is scarce for analyses of the underlying causes of true papilledema in this patient population. The authors identified 38 patients with papilledema from diagnostic codes and discovered that the most common etiologies were idiopathic intracranial hypertension (IIH) (42%), craniosynostoses (18%), and intracranial tumors (16%).[3] Together, these causes accounted for over 75% of cases of papilledema in children. Strikingly, the authors report that only 1 of the 38 patients had been referred specifically to rule out papilledema. The remaining cases included primary hydrocephalus and infectious and inflammatory etiologies, among others.[3]

This article highlights several concepts that are important for both residents and ophthalmologists practicing in community settings to consider. First, IIH accounts for nearly half of all cases of papilledema in this pediatric population. While headache and diplopia are the most common symptoms, over one-third of patients are asymptomatic.[3] In addition, the nature of headache symptoms varies widely in children with IIH where, for example, the headaches may be constant or episodic, and focal or diffuse.[4] Second, although the authors admit that patients with craniosynostosis may have been over-represented in their study due to local referral patterns, routine ophthalmic evaluation of these patients is important since none of them had presented with signs of elevated ICP. Third, the percentage of patients with papilledema due to an intracranial mass is not insignificant, even in an ambulatory clinic. As a result, urgent neuroimaging is prudent when papilledema is suspected.

Finally, although not specifically highlighted by the authors, over one-third of the patients in their study with papilledema had strabismus at presentation. A cranial nerve six palsy is the most common cranial nerve palsy associated with elevated ICP;[5] however, comitant esotropia, exotropia, and other cranial nerve palsies were also identified in this study.[3] This association reiterates the importance of a complete fundus examination, including specific attention to the optic nerves, in patients presenting acutely with either comitant or incomitant strabismus.

Referenced article: https://www.canadianjournalofophthalmology.ca/article/S0008-4182(18)31313-9/fulltext

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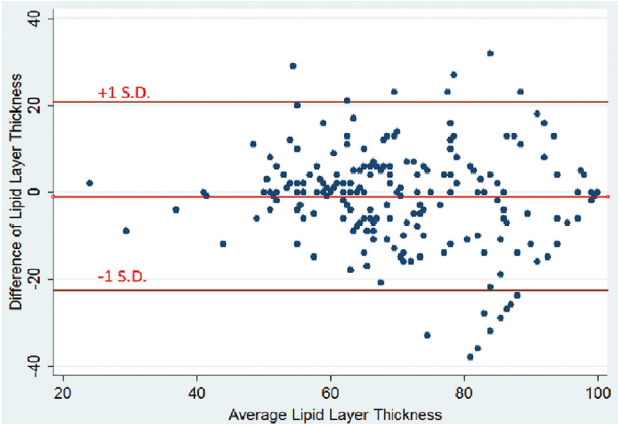

Value of lipid layer thickness and blinking pattern in approaching patients with dry eye symptoms

Jenny Qian, PGY-1

Dry eye is a common ocular surface disorder affecting millions of people worldwide.[1] Dry eye disease (DED) represents a heterogenous group of conditions associated with compromised ocular lubrication and can be categorized into two major forms: aqueous-deficient dry eye characterized by decreased tear secretion, and evaporative dry eye that results from increased tear evaporation.[1,2,3] Given the multifactorial nature of DED, diagnosis is often based on a combination of signs, symptoms, and clinical tests.[4] However, inconsistencies between reported symptoms, observed signs, and clinical test results can complicate the diagnosis.[5] In this issue, Chou et al. explore the relationship between dry eye symptoms and dry eye tests/parameters.[6]

Patients ≥ 20 years old with and without complaints of ocular surface irritation were recruited from the Taipei Veterans General Hospital. Dry eye symptoms were measured with the validated Standard Patient Evaluation of Eye Dryness (SPEED) and Ocular Surface Disease Index (OSDI) questionnaires. Dry eye parameters/tests included the tear film lipid layer thickness (LTT) and blinking patterns evaluated by the LipiView interferometer, tear film breakup time (TFBUT), and Schirmer I test. The ocular protection index (OPI), used to assess the effects of tear film instability, was determined based on the TFBUT and interblink interval.

A total of 115 patients (229 eyes) were enrolled in the study. Those with poor LLT signal and LLTm (mean LLT measurements from the same eye) ≥ 100 nm were excluded, resulting in 107 participants (203 eyes) included in analysis.

The authors found an inverse relationship between LLTm averaged between eyes (aLLTm) and SPEED/OSDI scores (p=0.0030, r=-0.284 and p=0.0054, r=-0.267, respectively). Clinically, patients with LLT ≤ 69 nm were more likely to have SPEED/OSDI scores ≥ 10 (sensitivity and specificity greater than 60%). These results are similar to those reported by Blackie et al, in which patients with DED had thinner LLT and among those with SPEED scores ≥ 10, 74% had LLT ≤ 60 nm.[7] Of note, many patients with clinically obvious meibomian gland disease (MGD) had LLT ≥ 100 nm and if these groups were included, the significance of the correlation between LLT and symptoms decreased. This indicates that both patients with thin LLT and LLT greater than 100 nm tend to be symptomatic. The authors also found an inverse relationship between LLTm and Schirmer I test (p=0.0002, r=-0.1857) — eyes with thinner LLT produced more aqueous tears, which may be a compensatory mechanism for tear film instability. Again, including patients with LLTm ≥ 100 nm decreased this correlation, which was also found by Eom et al.[8]

Many exogenous and endogenous factors affect blink rate, such as lighting, environmental irritants, ocular surface or systemic conditions, and fatigue;[9] as a result, DED patients can exhibit both higher or lower blinking rates. Expectedly, there was no correlation between average total blinking rate, complete blinking rate, incomplete blinking rate, or OPI with SPEED/OSDI scores. However, the authors did discover a significant correlation between SPEED/OSDI scores and average incomplete to complete (i/c) blinking ratios (p=0.0048, r=0.2706 and p=0.0234, r=0.2190, respectively). This suggests the nature of the blink is more significant than the blink rate itself for dry eye symptoms.

This study by Chou et al. demonstrates the utility of LLT measurements for the diagnosis of dry eye and the relationship between LLT and i/c blinking ratios with dry eye symptoms. Considering the study limitations, future exploration of how LLTs greater than 100 nm and how the quality of the lipid layer and other tear components correlate with dry eye symptoms is warranted.

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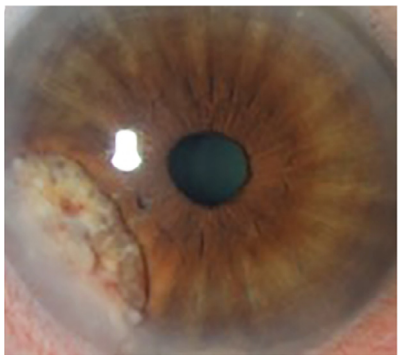

Fine-needle aspiration biopsy for suspected uveal metastases

Aishwarya Sundaram, PGY-4

Uveal tract is the most common ophthalmic location for hematogenous spread of metastatic tumors, which is thought to be secondary to the significant blood flow within the choroid.[1] Based on large cohort studies, the most common origin for uveal metastases were from breast (37%), followed by lung (26%), kidney (4%), gastrointestinal (GI) tract (4%), cutaneous melanoma (2%), lung carcinoid (2%), prostate (2%), thyroid (1%), pancreas (1%), other sites (3%), and unknown (16%).[1] Fine-needle aspiration biopsy (FNAB) provides a minimally invasive diagnostic tool for identification of a uveal metastasis, which can be helpful in identifying the primary source and for directing treatment decisions. In this issue, Bellerive and colleagues investigate the diagnostic accuracy of cytological analysis by comparing it with the subsequent clinical course in patients with nonmelanocytic intraocular masses.[2]

The authors conduced a retrospective review of 28 consecutive patients who underwent a diagnostic FNAB and recorded their age, sex, and primary malignancy origin site (if known).[2] The FNAB samples were obtained using a 25-gauge needle using a trans-corneal, trans-scleral or trans-vitreal approach. Previous studies have shown that 25-gauge FNAB can provide sufficient cellular aspirate for diagnostic accuracy in 79% of cases.[3] In the present study, the samples were reported based on cytological and immunocytochemical testing. The diagnostic report of the cytologic results were classified by the authors as positive, negative, atypical and non-diagnostic. The diagnosis was then correlated with the clinical course to determine the diagnostic accuracy.[2]

Of the 28 included patients, the aspirates were obtained from the following locations: 18 (64%) choroid, 4 (14%) iris, 3 (11%) ciliary body, 2 (7%) episclera and 1 (4%) subretinal space. Patients had a median follow up time of 13.5 months. Of the 28 FNABs, adequate samples were obtained in 25 (89%) cases; with 19 (68%) positive for metastasis, 4 (14%) negative biopsies and 2 (7%) atypical samples. Of the 19 patients with a positive biopsy, 16 (84%) patients were found to have uveal metastases: 9 adenocarcinomas, 3 small cell carcinomas, 3 non-small cell carcinomas, and 1 para-aortic paraganglioma of the iliac region. The other three patients with a positive cytology were found to have uveal lymphoma. All 19 patients were confirmed using clinical and radiologic confirmation to have an accurate diagnosis. Thus, the authors conclude that the overall specificity was 100% and sensitivity was 87.5% of patients. However, it is important to note that of the negative cases: 2 patients were later identified to have adenocarcinoma, 1 patient with *Cryptococcus neoformans* and another patient with nonspecific inflammation. Also, three of the nondiagnostic patients were later noted to have adenocarcinoma (1), extra-marginal zone lymphoma (1), and uveal schwannoma (1). Given the high rate of missed diagnoses in non-positive FNAB patients, the authors suggest that these patients should be closely followed and repeat FNAB as well as other clinical-radiologic investigations should be used to manage these patients.

While initially, this study may appear to be of limited significance due to its small sample size, it provides diagnostic confirmation by comparing the aspirate diagnosis with the patient's clinical course. Furthermore, the results are similar to a larger study that showed similar results of FNAB reliability in 159 suspected uveal metastasis patients,[4] but that study failed to compare diagnostic accuracy with clinical course and radiologic investigation.

Clinical practice point: FNAB is highly specific (100%) for the diagnosis of amelanotic uveal metastasis but the sensitivity of the test is only 87.5%. Furthermore, if a patient does not have a positive result after FNAB then they should be followed closely as they may still have an underlying malignancy or other diagnosis that may require treatment.

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