Introduction
History of the DES/OMS Abingdon Meetings
Remembering Michael Pranzatelli M.D.

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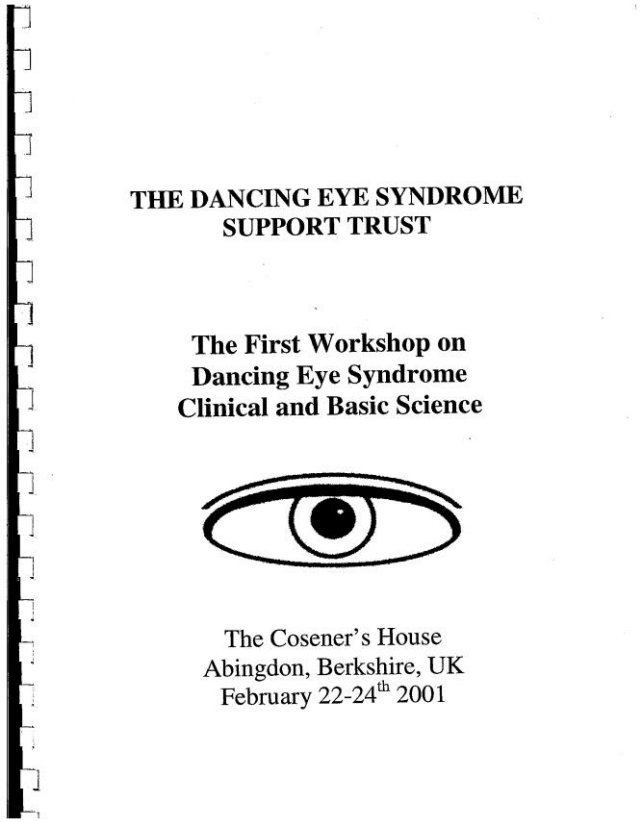
Introduction: my topics today

- Welcome (finally) to the postponed 2020, 2021, spring 2022 and finally, October 2022 10th DES/OMS Abingdon symposium
- Rather than a general overview of OMS, I will discuss the history of this meeting
  - No introductory video on a child with OMS today!

- Memories of and praise for Mike Prazatelli

2001 Abingdon symposium: How it all began



THE DANCING EYE SYNDROME
SUPPORT TRUST

The First Workshop on
Dancing Eye Syndrome
Clinical and Basic Science

The Cosener's House
Abingdon, Berkshire, UK
February 22-24th 2001

- The first DES workshop in 2001 was organized by Dr. Peter Beverly and colleagues and parents and families in the DES Support Trust
- Dr. Beverly had experienced a similar rare disease focused meeting combining families, clinicians and researchers and proposed this (unique) structure
- Jane Stanton-Roberts played a major role in organizing this and subsequent meetings
- The majority of attendees were from UK, a few from Europe and USA. Parents were primarily from UK.

2001: a beginning

- Clinicians and parents educating scientists about what is OMS
  - Mike Pike led off with his famous video of an OMS/DES child
  - 7 clinicians and 3 parents (although other parents attended and participated)
- Researchers: becoming interested in a puzzling disease, new to them but potentially related to their work:
  - Neuroimmunologists
  - Research neuroimaging
  - Oncologists
  - Basic scientists

Of speakers, 17 from UK, 5 from US, 1 from EU
We learned from each other, made new research contacts, and generated interest in research in DES/OMS

Subsequent sessions 2003, 2005, 2008, 2010, 2012, 2014, 2016, 2018:

Similar format, more diverse group of clinicians, scientists and parents

- More parents participated and have become active, ongoing participants
- Joclin Murphy 2008
  - Instrumental in writing parent/teacher/therapist guides
- Mike Michaelis 2012
  - OMSLife became meeting cosponsor as of 2016
- Ian Grummitt 2018
  - steering committee as of 2018

- A wider group of clinicians including pediatric oncologists from USA, UK and EU
- Basic scientists, statisticians, neuropsychologists
  - More about outcome
  - Development of databases
  - More discussion of treatments
  - More interest in adult survivors of OMS

A few notable landmarks:

B-cells and cytokines in CSF

- Rituximab proposed, begins to be used with more intensive treatment

COG randomized clinical trial:

2004-2013, presented at 2016 meeting

European clinical trial and database

OMSlife database, NORD sponsorship

Pavlov grants for neuroblastoma specify funds for OMS

The long-awaited consensus paper is finally published

2022

POOMAS international database

2019-present

Some ongoing challenges:
A partial listing

- No identification of antigen and antibody in OMS
  - No available surrogate bio-marker for OMS disease activity (i.e. serum test), despite some initially promising hints
- Lack of full consensus on best initial treatments, best treatment for relapses, treatment of refractory OMS
  - Absence of randomized clinical trial data other than COG trial
- How to improve cognitive outcomes, prevent learning disabilities
  - Are post-OMS learning disabilities qualitatively different from "common" types of dyslexia, ADHD? Is treatment different?
- Late sequelae including anxiety disorders, other psychiatric problems
  - Are very late relapses (such as with pregnancy) the same process?
- Finding ongoing specialty care for adults who are OMS survivors

Michael Pranzatelli M.D.

- Michael Pranzatelli MD devoted most of his career to the study of OMS.
  - He saw virtually no other patients for at least the last 15 years of his practice.
  - He died of pancreatic cancer in fall 2018
  - He attended this meeting first in 2001, last in 2016, but not in 2018

Initial interest in neuropharmacology of myoclonus lead him to OMS

- He was a neuropharmacology fellow at CHLA from 1982-85
  - Although he and I were both there in those years we had minimal interaction as he was primarily in bench lab without clinical involvement
- He joined faculty at Columbia University School of Medicine in 1985, moved to GW school of Medicine in 1989, then to University of Southern Illinois, where he established the National Pediatric Myoclonus Center. He left SIU in 2014 and moved to Florida where he had a private practice devoted to OMS. He was on the faculty of University of Central Florida College of Medicine.
- Through all of this, he leaned heavily on his wife, Elizabeth Tate, a nurse-practitioner, who did a great deal of the patient management (i.e. parent management) as well as collaborated in his research.

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Various accomplishments regarding OMS:

- Described activated lymphocytes (CD19+) in CSF of children with active OMS
- Reported prevalence of oligoclonal bands in CSF correlated with disease activity
- Described presence of a number of cytokines and chemokines in CSF of children with OMS
- Introduced rituximab as a major component of OMS treatment
- Attempted to standardize aggressive “first line” treatment of OMS, advocating for combination of ACTH, rituximab and IVIG
- Launched OMSUSA.org in 2001, the first comprehensive OMS website for both parents and physicians
- Published 152 manuscripts, at least 53 specifically on OMS

First publication specific to OMS:

The neurobiology of the opsoclonus-myoclonus syndrome. Pranzatelli MR; *Clinical Neuropharmacology. 15(3):186-228, 1992 Jun.*

Published in 2018
(while on chemo for cancer)

- 11 publications, all but one first author, all but one regarding OMS: Notable among them:
  - Michael R Pranzatelli [1], Elizabeth D Tate [1], Nathan R McGee [1] Multifactorial analysis of opsoclonus-myoclonus syndrome etiology ("Tumor" vs. "No tumor") in a cohort of 356 US children 2018 Pediatr Blood Cancer,. Aug;65(8):e27097

  - Michael R Pranzatelli [1], Nathan R McGee [2], Elizabeth D Tate [3] Relation of intrathecal oligoclonal band production to inflammatory mediator and immunotherapy response in 208 children with OMS J Neuroimmunology 2018 Aug 15;321:150-156.

Thanks and final word

- We all owe a debt to Michael Pranzatelli, Elizabeth Tate, and the scores of OMS families to travelled to his centers and contributed to his research.

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Thank you!