Pediatric Neurology 90 (2019) 3–7

Contents lists available at ScienceDirect

Pediatric Neurology

journal homepage: www.elsevier.com/locate/pnu

Perspectives in Pediatric Neurology

What We Need to Know?

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ARTICLE INFO

Article history:
Received 30 July 2018
Accepted 31 July 2018

Keywords:
Neurodevelopmental disabilities
Pediatric neurology
William Osler
Cerebral palsy
Training

"*The value of experience is not in seeing much, but in seeing wisely.*"[1]

Sir William Osler

Introduction

The terms of reference of the Hower Award describes it as "a prestigious award to honor a child neurologist and member of the Child Neurology Society who is highly regarded as an outstanding teacher and scholar, and additionally has given a high level of service to the Child Neurology Society (CNS). Particular emphasis is placed on contributions to child neurology at other national and international levels."[2] To be designated a recipient is to be both honored and humbled, as nothing is sweeter than the recognition of one's peers that a professional journey and career has not only be well-spent, but deemed especially meritorious.

Probably the most recognizable outward attribute of the Hower Award is the lecture given by the awardee as a plenary address on the final day of the CNS annual meeting. The lecture at the CNS typically falls into two broad categories of content and style. The awardee either gives a talk that is a summation of a personal scientific journey focusing in depth on a particular topic, or it is one that is philosophical, expansive, reflective, and at times whimsical. The possibility of a pulpit free of content constraint is too tempting an offer for me to not indulge the latter option. A fellow McGillian, Sir William Osler, will join us. Osler was never at a loss for prescient words, and quotes from his writings will be used to frame my presentation.

What we need to know?

The expanse and length of the textbooks of our field (e.g. Swaiman's, Volpe's, Aicardi's etc.)[3-5] is eloquent testimony to the breadth of factual knowledge necessary to the practice of child neurology. To truly keep up, one has to utilize powerful search engines and computing power that are now consulted on almost a daily basis.

What I hope to convey in this paper are not facts, but "truisms" that are enduring pieces of wisdom derived from my experience over time. Any child neurologist who has trained with me has endured these "truisms" in one form or

This article is based on the Hower Award Lecture given at the 43rd Annual Meeting of the Child Neurology Society in Columbus, Ohio in October 2014.

Conflict of interest and source of funding statement: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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https://doi.org/10.1016/j.pediatrneurol.2018.07.017

another on multiple occasions (repetition always drives the point home). These "truisms" for me constitute the essential core of successful practice that has stood the test of time.

Why neurology?

1. *The problem of neurology is to understand man himself.* - Wilder Penfield[6]
2. *To understand the man, you have to start with the child and family.* - Michael Shevell (MS)[7]
3. *Remember there is a neuro in neurodevelopmental.* - MS
4. *And remember too there is a developmental in neurodevelopmental.* - MS
5. *While you will not 'cure' much (if anything), you can always leave the child and family a bit better off than when you first met them.* - MS

The quote from Penfield is literally chiseled into the concrete exterior wall approaching the entrance of the Montreal Neurological Institute. It enables us to reach for the heavens. It makes us fallible. It makes us the individual each of us is. It enables us to experience emotions. And we die irreversibly when our brain ceases to function. I suspect at root this is the reason we each chose this specialty in the first place. And it all begins in childhood in the context of a family. That is the beginning of the story and it greatly determines how the chapters of an individual's life will play out.

Neurodisabilities (global developmental delay, intellectual disability, autism spectrum disorder, and cerebral palsy) collectively are a major clinical challenge, are brain-based entities, and progress in these disorders will only come from understanding the brain.[7] Yet these disorders arise against the backdrop of a maturing brain, leading to challenges that evolve across the lifespan. However, the brain is a harsh mistress. We know too little to cure much and indeed this aspect unfortunately has often been used by insensitive preceptors to dissuade the bright student from pursuing a career path in the clinical neurosciences. However, we can always in some way improve things for our patients and their families.

Diagnosis

"*One finger in the throat and one in the rectum makes a good diagnostician.*" - Sir William Osler[8]

6. "*Always measure the head circumference.*" - MS
7. "*The physical examination begins the moment you first see the child and continues throughout the encounter with the child and family. If at all possible, watch the child play.*" - MS
8. "*Diagnosis is about history and physical examination. Everything else is confirmatory.*" - MS
9. "*Never call a child dysmorphic until you have seen both parents.*" - MS
10. "*Cannot measure it, but curiosity is the most reassuring thing one can observe on the examination of the infant or child.*" - MS
11. "*MD stands for Medical Detective. Families need to know why there is something 'wrong' with their child.*" - MS
12. "*The earlier the diagnosis, the earlier the intervention, the better the outcome, however or whatever measured.*" - Annette Majnemer, PhD, OT, FCAHS

These observations are about the fundamentals. Yes, we are smarter now due to technological advances, especially in the domains of imaging and molecular diagnostics, but it all begins with something that Sir William would be comfortable with. Measuring the head circumference is the most overlooked and under-appreciated segment of the physical examination in pediatric neurology. Not only do we need to measure it, but also plot it. Children are naturally averse to strangers. Yet we can learn so much before the formal "hands-on" portion of the examination where we risk losing their cooperation. I always take the time to go to the waiting room to call the child and family in. Watching the child walk gives so many data points to informing the examination. Giving the child an opportunity to play while speaking to the family allows not only for detailed observation, but an opportunity to be informed about developmental levels, behavior, and temperament.[9] Seeing curiosity expressed in how the young child seeks to explore their surroundings provides reassurance about cognition and social connection. Diagnostic testing is never random and must always be informed by what we suspect is going on. Do not order a test because you think something might be going on, order it because you know something is happening.[10] Dysmorphology is indeed in the eye of the beholder and in a multicultural society we can not jump to conclusions. And diagnosis is the essential precondition for what follows. We need to ask, and answer, a very pointed question: why this child? If we answer the question correctly, the answer is the key that unlocks the right doors going forward regarding treatment, interventions, prognosis, and recurrence.

Outcomes research can be summarized in the last quote above. There are no exceptions to this observation I can find. It also gives us the imperative to do what we can to move forward in time the diagnosis of pediatric neurological disorders so that we can continue to improve outcomes.

Caring

"*The good physician treats the disease; the great physician treats the patient who has the disease.*" - Sir William Osler[11]

Sir William was talking about physicianship and family centered care a century before anyone else was.

13. "*Every family is different and dysfunctional in its own way.*" - MS

14. "*Ask the magic wand question: You will often be surprised by the answer.*" - MS

15. "*Focus on what you can change to make things better for the child and family.*" - MS

16. "*Pediatric neurology is a team sport and there is no letter 'i' in team.*" - MS

17. "*Disability is a simple equation: Disability = Impairment [Biology] + Barriers [Environment].*" - MS

18. "*If the family believes in the priest, minister, rabbi, imam, shaman or witch doctor: Perhaps we should too.*" - Meaghan Shevell BA, MA

The first quote owes itself to Tolstoy and the opening line of his epic novel Anna Karenina. Context is both our challenge *and* our opportunity. Every child and family is different. Cookie cutter approaches to care are doomed to failure. We need to engage with families as partners in care and listen to what matters to them. We have to move away from stubbornly focusing on "fixing the brain" (something we often cannot do) to rather focusing on what they want us to address and help with. Indeed this will likely be something that we can change. And in changing, we will leave the child and family better off and we will be more satisfied with our professional efforts. Ultimately, it is not about eliminating the child's quadriparesis, but perhaps it may be about a good night sleep, or an easier diaper change, or less pain. Hence the "magic wand" question: if we could fix one thing, what should we focus our efforts on? In the disability equation, of the two variables, it is the barriers (environmental, societal, cultural, and educational) that are most amenable to change. Maximizing participation is a goal we can all agree on. And the good news is that we are not alone in this effort. Effective partnerships with other health professionals leverage our skill set to maximum effect to the benefit of the child.

The last quote reinforces the contextual challenge we face and the need to make therapeutic liaisons with the families we treat. It also demonstrates how valuable the social sciences are in the proper practice of medicine.

Prediction

> "*Medicine is a science of uncertainty and an art of probability.*" - Sir William Osler[12]

I find this quote most aptly summarizes the daily practice of academic child neurology. We are continually on a shifting surface of what we know, trying to be an oracle of what will be. Often the information available is incomplete, adding to our challenge. Often the questions asked are not those that we can answer, so we spend our efforts frequently answering questions that are not even posed.[13]

19. "*Any organ system can bring a child into a NICU or PICU: However important long term morbidity will invariably include something neurological in origin.*" - MS

20. "*The most important test in neurology is precious time.*" - MS

21. "*Except in an emergency situation, time is your most important diagnostic, therapeutic and prognostic ally.*" - MS

22. "*Parents have but one child before you: The only probability statistic that matters to them is 0 or 100%.*" - MS

23. "*The one consistent risk factor for any disorder or outcome is low socioeconomic status or social vulnerability.*" - MS

24. "*The most important, perhaps the only, prognostic question that really matters is: Will this child and family be happy? And it's the question you cannot answer.*" - MS

In the NICU and PICU we have still not yet mastered the capability of moment-to-moment monitoring of brain function. So much of what our esteemed colleagues encounter in these technology intensive milieus with fragile newborns and children is potentially deleterious to a susceptible brain. Neurocritical care is the future of critical care. The privilege that observation and time offers enables the refinement of precision. Resiliency, recovery, and plasticity are all so intrinsically idiosyncratic that inferring from the general to the particular is fraught with inherent unavoidable error. We talk in percentages to stressed parents knowing full well that for them it is an "either/or" roll of the dice. And we talk about things that we know, avoiding the important questions of quality of life and happiness that are so contextually dependent. Some of the happiest children are among the most objectively "disabled" children. And like an early diagnosis and outcome, low socioeconomic status and social vulnerability are potent modifiers of both causation and outcome. Epigenetics is providing the science to explain this, but this observation in wealthy nations such as ours is a clarion call for social justice and improved social and health policy. A rising tide should raise all, not just some, ships in the harbor beyond the rocks.

Science (and heart)

> "*To study the phenomena of disease without books is to sail an uncharted sea, while to study books without patients is not to go to sea at all.*" - Sir William Osler[14]

Sir William was the master of bedside teaching and the original clinician-scientist. The bedside was his arena, extrapolating in both directions to and from the bench. We learn from our patients and we learn from those toiling away in the lab. Fortunate are we to be the conduit between the two.

25. "*To know cerebral palsy is to know pediatric neurology.*" - MS

26. "*Every clinical encounter represents a teaching and research opportunity.*" - MS

27. "*Sometimes regretfully no brain is better than bad brain.*" - MS

28. "*Pediatric neurology is transitioning from a descriptive discipline to an interventional one: Our time is now.*" - MS

29. "*Words of comfort are sometimes the only therapeutic intervention we can offer: You have to recognize and accept this when it occurs.*" - MS

30. "*The mind thinks it and the brain does it. And the mind is in the brain.*" - Allison Shevell BSc, MDCM, FRCP

Cerebral palsy really does encapsulate all that transpires in pediatric neurology.[15] If there is one chapter in the textbook to know wholly, this is it. Heterogeneous presentations, causes and outcomes, findings on neurological examination, anatomic correlations evident on imaging and pathology, comorbidities across the spectrum including epilepsies, cognitive limitations, behavioral challenges, sensory impairments, emerging genetic implications, sleep disturbances, movement disorders, feeding issues, and a chronic course across the lifespan. It also offers a range of therapeutic interventions and partnerships to optimize outcomes with multiple other health professionals. The heterogeneity of child neurology makes every case we encounter a teaching opportunity. The rarity of many disorders within a single practice, together with the aforementioned heterogeneity, suggests that research opportunities are plentiful, but unfortunately strikingly under-utilized. Perhaps the widespread utilization of electronic medical records platforms and the emergence of Big Data will improve this situation. Exquisitely localizing and removing the bad brain is the *sine qua non* of the miracle that is epilepsy surgery. Therapeutic hypothermia, transcranial magnetic stimulation, intravenous immunoglobulins, exon skipping therapies for SMA, and the promise of CRISPR technology all demonstrate the paradigm shift that is occurring before us. Our time is indeed now, as increasingly we are enabled to intervene in the pathogenesis and evolution of neurological disorders. However, sadly this is not true for all afflicted and all too frequently, we reach our limits in what we can actively do. In such situations we must remember that palliative care is not "no care". There is still enormous power in simply being there to comfort and to provide solace and support.

There has often been a tangled relationship between a philosophical construct (mind) and a tangible reality (brain). This dichotomy and the problem it creates is entirely of our own creation. Our minds are our brains. The complexity of the underlying machinery likely precludes us from fully understanding this relationship, but it is sufficient to know that disorders of the mind have a physical basis that is influenced by environmental and contextual circumstances.

Work and Life

"*Without egotism and full of feeling, laughter is the music of life.*" - Sir William Osler[16]

It seems in this quote that Sir William beat the work-life integration and physician wellness proponents by a century.

31. "*Look good, feel good, do good!*" - MS

32. "*You are spending your days with children: Have fun. Make them laugh and you will smile too.*" - MS

Pediatric neurologists have many difficult challenges and deal with truly devastating cases. Laughter helps us, and the children and families we care for, deal with this oppression. It is rare that a patient encounter does not include an elicited smile, chuckle, or a hearty laugh. We can do good so much more readily if we too feel good ourselves. It is so important that we have balance in our lives; spending time with people we care about, engaging in activities and pastimes that give us pleasure. And for those of us vested in this wonderful field, nothing should give us a greater sense of joy then making a child laugh in delight.

Conclusion

"*One special advantage of the skeptical attitude of mind is that a man is never vexed to find that after all he has been in the wrong.*" - Sir William Osler[12]

The "truisms" quoted and explored above are but propositions for discussion and reflection. They are by no means exhaustive or complete. They are but a part of what we need to know as child neurologists. A useful core on what is no doubt a long, winding and never-ending road to providing care and comfort.

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