

What are the prevalent neurological conditions treated by the paediatric department at Columbia? Does this reflect disease patterns worldwide?

In my time at CHONY I saw a variety of neurological presentations, spanning vascular, traumatic, demyelinating, metabolic, autoimmune and genetic disease. However, the most prevalent condition seen and treated on the floor was epilepsy. I was able to see a vast range of epilepsy syndromes including infantile spasms, benign rolandic epilepsy, a possible case of panayiotopoulos syndrome, juvenile myoclonic epilepsy and presentations mimicking seizures; non-epileptic attacks. All cases were discussed in detail with Dr Sultan during ward rounds and with the epilepsy team. We were also lucky enough to have teaching opportunities with senior figures in paediatric neurology, from case discussions with Dr Riviello and neurosurgeon Dr Feldstein to afternoon teaching with Dr DeVivo, who described GLUT1 deficiency.

As a specialist centre in one of the leading children's hospitals in the world, with an attached epilepsy monitoring unit, it was unsurprising to be seeing children with complex epilepsy syndromes and hard to control seizures. However, it is also unsurprising from a worldwide perspective - epilepsy is one of the most prevalent neurological disorders, and at least 50% of cases begin in childhood. The estimated proportion of the general population with active epilepsy is between 4-10 per 1000 according to the WHO, who also report that epilepsy tops the list of diseases most frequently seen by a neurologist. Furthermore, symptomatic epilepsy, which is secondary to brain damage in the prenatal or perinatal period, congenital abnormalities or head trauma is far more likely in a population of children with other neurological disease. For example, one of our patients with non-accidental injury presented with myoclonic jerks, likely to represent epileptic events. Many of our patients had autism and concomitant seizures, which is common in this population.

How does the organization of paediatric services in the US differ from the UK?

Unlike in the UK, children in the US see a paediatrician rather than a general practitioner for their primary medical care. They attend the paediatrician regularly for well child checks, not a routine part of care in the UK. They would then be referred to a specialist paediatrician for management of more complex disease, such as neurology. In the UK, children tend to see specialists less often, as paediatricians are already considered specialists. As a consequence, children with neurological disease in the US seem to undergo more detailed investigations. I saw cases where these led to incidental findings such as microadenomas of the pituitary, which were unlikely to contribute to the presentation. However, in journal club we discussed a paper showing that mortality from brain tumours is higher in the UK than the US, which is likely to be a benefit of this more aggressive approach to investigation.

In terms of training, I was fascinated to find out that newly qualified doctors could apply straight to a residency program in paediatric neurology. Such specialization so early on in a medical career, in contrast to our programmes in the UK, has many benefits. The residents I worked with were already extremely knowledgeable in their field, and highly competent in history taking, examination and medical decision-making. They were able to take care of many issues on the floor or arising from consults. While on a personal level it forces students to make big decisions very early on, I think in general it makes for a high standard of medical care.

Discuss an interesting case seen in paediatric neurology, from presentation to outcome.

A 7-year-old boy presented with a 48 hour history of acute onset continuous left-sided perioral and facial twitching, sometimes involving the eyebrow and occasionally the right side of his face. It was not associated with any change in conscious level, jerking of the limbs, incontinence or tongue biting. However, he was unable to eat due to the local discomfort. He had no weakness, sensory disturbance, hearing loss or other neurological deficit. There were no systemic symptoms.

His previous medical history included a prenatally diagnosed posterior fossa arachnoid cyst, which had been fenestrated at age 2. He also had meningitis at 3 months, when he had a 2 week course of intravenous antibiotics. Finally, 22 months he was diagnosed with autism, having delayed speech and impaired interactions. At the time of presentation he had minimal language and communicated predominantly nonverbally.

On examination the facial twitch was prominent, but there were no altered mental status, cranial nerve abnormalities, limb weakness or motor signs in the arms or legs. He would not participate in sensory exam.

His basic labs were unremarkable. A more specific workup for autoimmune disease was negative, with the serum NMDA Ab negative. LP was negative for HSV, VZV, CMV, enterovirus, and autoantibodies but had 2 oligoclonal bands. EEG showed epilepsy partialis continua. MRI imaging showed no acute abnormality, and the posterior fossa cyst was thought to be comparable to a study from 2010.

On admission to hospital he was treated initially with lorazepam and loaded with phenytoin. He continued maintenance phenytoin 70mg bd. On day 3 he was started on clonazepam 0.25mg and levetiracetam 1500mg bd, to which the symptoms were unresponsive. With increasing suspicion for an autoimmune cause, he had a 5 day pulsed high dose IV prednisolone 20mg/kg, which was then switched to oral prednisolone and three rounds of plasmapheresis. The symptoms appeared to lessen at night, and he was discharged 2 weeks after admission with NG tube in situ.

This case illustrated well the complexity of cases that the team would see – and also the approach, which emphasised utilising the latest research findings to inform patient care. Unfortunately it was also a reminder that despite the best efforts and contemporary tests, we cannot always reach a diagnosis for patients in this time frame.

Is paediatric neurology an area I would consider as a future career?

My experiences on elective at CHONY have further clarified for me that my future lies in neurology. I took full advantage of the incredible learning opportunities on offer every day in both the institute of neurology and within the department. The close links between paediatric neurology and the adult service allowed me to attend conferences on deep brain stimulation, autonomic neuropathy and stem cell therapy for amyotrophic lateral sclerosis. Morning report was a highlight every week, where the patient case was discussed, from initial presentation to diagnosis – bibrachial diplegia, the newly-described DPPX encephalitis and autoimmune demyelinating encephalomyelitis.

I have also really enjoyed the opportunity to understand how neurological disease manifests at different ages, and how to examine for weakness, sensory loss, cranial nerve signs and cognition in babies, toddlers and adolescents. I enjoy working with children and the US paediatric neurology programme would be great for me. However, the UK system, where I would have to commit to 5 years of general paediatrics after 2 years in the foundation programme before specialising in the field would require a long period away from my core interest of neurology.

However, within the field of neurology there are likely to be opportunities to see paediatric patients, and this is certainly something I will research for the future.