Consultant's Corner

Practical Answers To Your Everyday Questions

Cerebellar Degeneration Treatments

1.

What are the causes of cerebellar degeneration and how do you treat and manage it?

Question submitted by:
Dr. Paul K. Chiu
Edmonton, Alberta

A larger variety of hereditary and acquired disorders can result in progressive loss of cerebellar function. I will focus my discussion here only on cerebellar degenerative disorders that present insidiously and are genetically determined. Most autosomal recessive cerebellar ataxia present early in life usually before 20-years-of-age. Autosomal dominant ataxia usually present later in life. Friedreich's ataxia is the most common autosomal recessive ataxia presenting with limb ataxia, dysarthria, areflexia and peripheral neuropathy. Electrocardiographic abnormalities are frequent and require careful monitoring. Cardiac arrhythmias or heart failure may require treatment. Other recessive cerebellar ataxias may have associated gonadal deficiency that may require hormonal replacement. A number of late onset autosomal dominant ataxias have been described. The cerebellar dysfunction is associated with prominent involvement of the spinal cord and the brainstem. Autonomic dysfunction may be seen in some patients. BP fluctuations (frequently postural hypotension), bradycardia or gastric paresis may require treatment that in most cases is symptomatic. If there is prominent spasticity, the use of antispasmodic medications may improve function and improve pain control. There is no known treatment to slow down progression of the underlying cerebral ataxia.

Resource

1. Harding AE. Cerebrellar and Spinocerebellar Disporders. Neurology in Clinical Practice. Editors Bradley WG, Daroff RB, Fenchel GM, Mardsen CD. 1996 Pages 1773-1792 Butterworth-Heinemann

Answered by:

Dr. Ashfaq Shuaib

Brown Pigmentation of the Skin's Surface

2.

What medical conditions cause brown pigmentation of the face, arm and body surface?

Question submitted by:
Dr. Dominic Eustace
Riverside Estates,
Saskatchewan

Causes include Addison's disease, Wilson's disease, scleroderma, hemochromatosis, malignancies (especially lung) and metastatic melanoma. Occasionally, advanced HIV infection, generalized acanthosis nigricans and neurofibromatosis can lead to a generalized darkening of the skin.

Answered by:

Dr. Scott Murray

Diagnosis and Treatment of Gastroesophageal Reflux in Infants

3.

Can you please comment on the diagnosis and treatment of gastroesophageal reflux disease (GERD) in infants?

Question submitted by:
Dr. Virginia Keeping
Mount Pearl, Newfoundland

The reflux of gastric contents into the esophagus or indeed all the way up to the oral cavity is very common in infants and in the absence of symptoms probably should be neither investigated or treated, as this is a developmentally normal event that will resolve over time such that by 18 months reflux becomes quite uncommon. If, however, reflux is accompanied by symptoms such as failure to thrive, respiratory complications or difficulty with feeding, this indicates the presence of GERD, which may indeed require intervention. The diagnosis begins with clinical suspicion and a careful history and examination is the first step in diagnosis. Endoscopy may be of benefit, but esophageal pH studies and upper GI radiological studies are of limited value, in that many normal infants with have reflux without GERD. In the event that the child has symptoms and a clinical diagnosis of GERD is made, a trial of lifestyle modifications can be undertaken, which may include thickening feeds or upright posture after feeding. Thickening is typically done with rice cereal. It should be noted that the evidence to date suggests that these modifications may alter the degree of reflux but there is no evidence that they change reflux-related pathology. These lifestyle changes should also include avoidance of tobacco smoke exposure. If there is a smoker in the home, the smoker should probably quit or at the very least only smoke outdoors. It should also be emphasized that these changes do not include sleeping in the prone position, due to the risk for sudden infant death syndrome. If the child has been losing weight, in addition to lifestyle modifications, there needs to be a careful assessment for other conditions such as celiac disease or dietary protein intolerance. If conservative measures do not produce improvement, then consideration for a trial of acid-reduction therapy should be given. The PPIs are probably the drugs best suited for this trial, but it cannot be over-emphasized that this should only be after a trial of lifestyle modification, as evidence to date suggest that these drugs are over-prescribed to children. Omeprazole, lansoprazole, esomeprazole and pantoprazole have all been evaluated for use in children. An advantage of the PPIs over histamine type 2 receptor antagonists is that the PPIs do have reduced efficacy with prolonged use. There is essentially no role for prokinetic drugs in the management of most cases of GERD related to safety concerns and a specialist in the management of GI disease in children should manage children with GERD for whom a trial of prokinetic agents is being contemplated.

Answered by:

Dr. Michael Rieder

MRI Screening for Fibrocystic Breast

4.

Is there any value in MRI screening of a fibrocystic breast?

Question submitted by:
Dr. Teresa Cordoni
Port Coquitlam,
British Columbia

MRI screening is offered to women at high risk of breast cancer (BC) such as those BRCA1 or BRCA2 gene positive, or who have a strong family history of BC. MRI is very effective at delineating small lesions, particularly in dense breasts, a risk factor for BC, but mammography should still be done as some BCs are not detected by MRI. MRI does not show micro-calcifications associated with ductal carcinoma *in situ* as seen on mammography. Cystic lesions are well outlined with ultrasound which is a good adjunct to mammography and clinical exam. Fibrocystic breasts may be tender and swollen but are not, however, associated with increased risk of BC. Thus, while MRI can image fibrocystic breast disease very accurately, it is not suggested for this purpose at this time in most jurisdictions.

Answered by:

Dr. Cathy Popadiuk and Dr. Connie Hapgood

Recommended Biopsy for Patients on Warfarin

5.

What type of biopsy method would be recommended for a patient on warfarin?

Question submitted by:
Dr. Barb Secter
Edmonton, Alberta

More than two million patients in North America who take warfarin face a major problem should they need surgery or an invasive procedure. It is best to work with the specialist who is going to do the procedure to determine the proper pre- and post-procedure management of warfarin. In general, some procedures, such as some ophthalmic, endoscopic and dermatologic procedures, entail a low risk of bleeding and do not require that warfarin therapy be interrupted. If warfarin is withheld for five days, the INR usually falls to < 1.5 and surgery is usually safe. The need for bridging therapy (using heparin) depends on the patient's calculated risk of thromboembolism without it, the risk of bleeding with it and other factors. Anticoagulation therapy should usually be restarted on the day after the procedure.

Answered by:

Dr. Chi-Ming Chow

Measles, Mumps and Rubella Vaccine

6. If the first year measles, mumps and rubella (MMR) vaccine "takes" with a characteristic exanthem, is there need for the booster?

Question submitted by:
Dr. Santosh Paikatt
Cambridge, Ontario

Vaccination can "fail" in one of two ways:

1) Primary vaccine failure occurs when the vaccine is administered but no immune response is elicited. With live attenuated virus vaccines like MMR, this most often happens when the product has been mishandled (*e.g.,* break in the cold-chain) or when it is administered too early, in the presence of maternal antibodies
2) Secondary vaccine failure occurs when the vaccinated individual initially responds but loses the response (and protection) over time. It was long thought that the 4% to 5% primary vaccine failure rate was the only reason to receive a second dose of measles vaccine.

There is now good evidence that, in the absence of periodic boosting from wild-type virus, secondary vaccine failures can occur following MMR. Such fading immunity likely contributed to the measles outbreak in Eastern Canada in the early 1990s and to the on-going mumps outbreak in Canada in 2007 to 2008. It is not known if the small subset of children who develop mild symptoms following MMR (a clinical "take") have more durable immunity than those who have no obvious symptoms.

Answered by:

Dr. Michael Libman

Repeating Blood Tests for Iron Deficiency Anemia

7.

How often should you repeat complete blood count (CBC) in someone with iron deficiency anemia (IDA) on treatment?

Question submitted by:
Dr. Deanna Field
Truro, Nova Scotia

Typically, a patient with IDA who responds to treatment will have a reticulocytosis first around days four to seven followed by a hemoglobin response as early as one to two weeks. However, it may take several months for the anemia to resolve. Hence, it is not necessary in an otherwise stable patient to perform excessive testing. Although there are no specific guidelines for the monitoring of response to treatment, we routinely request CBC and serum ferritin every three months after initiation of therapy. Once the anemia is corrected, the underlying cause of iron deficiency is identified and no further iron loss is present, supplementation should be continued for at least another three to six months to replete the body's iron stores.

Answered by:

Dr. Kang Howson-Jan and Dr. Cyrus Hsia

Management of Ingrown Toenails

8.

What are practical management options for ingrown toenails in teenagers?

Question submitted by:
Dr. Anna Chlebak
Langley, British Columbia

The main problem leading to ingrown toenails is the lateral edge of the toenail growing into the periungual skin, leading to pain and secondary infection. This may result from the nail being cut too short at the lateral edge, as well as being worsened by repetitive pressure and the presence of thick periungual tissue.

In the early stages of inflammation, you can try non-surgical management. Treatment can include gently dislodging the lateral edge of the embedded nail plate from the inflamed nail fold, followed by placement of gauze or cotton under the sharp corner of the nail. Warm soaks can be done several times daily until the inflammation has cleared. Care should be taken during walking and exercise to minimize pressure on the toe and the nail should be trimmed with squared corners that extend distal to the end of the nail fold. Any infection/cellulitis can be treated with topical or systemic antibiotics especially in the diabetic or immunocompromised. The ultimate remedy is lateral nail plate excision with matrixectomy (usually with phenol) to ensure that the lateral nail plate does not regrow.

Answered by:

Dr. Scott Murray

Croup in Children

9.

What are the causes, symptoms and treatments for croup in children today?

Question submitted by:
Dr. Danielle Fisch
Canton-de-Hatley, Quebec

Croup is an acute respiratory disease in children characterized by barking cough and inspiratory stridor, often accompanied by vocal changes such as hoarseness. The etiological agent responsible for croup is most commonly parainfluenza type I, although a variety of other viruses can cause croup as well. Croup typically presents with a coryzal prodrome followed in a day or two by fever and barking cough associated with respiratory stridor. This is most pronounced at night and worsens for the next several days before improving. The usual management for croup includes fluids, antipyretics and humidity in the room, all of which are primarily for symptomatic relief. In the case of a child with significant upper airway compromise, inhaled adrenergic therapy with an agent such as L-epinephrine is indicated, administered as 0.5 mL/kg per dose (maximum of 5 mL) of a 1:1,000 dilution. It is given via nebulizer over 15 minutes. The major change over the past several decades in the therapy of croup has been the recognition that early use of steroids is associated with a significant decrease in the severity and duration of symptoms. This work, pioneered by Dr. T. Klassen from Canada, demonstrated that the use of a single dose of 0.6 mg/kg of dexamethasone given early in the course of croup had a major impact on how long children were symptomatic and how many required interventions such as hospital admission. Subsequent trials have demonstrated that this same effect can be provided by a dose of 0.3 mg/kg, with one trial showing positive results with a dose of 0.15 mg/kg. There is the most evidence for the use of a single dose of dexamethasone at 0.3 mg/kg, given early in the course of the disease.

Answered by:

Dr. Michael Rieder

Treating Attention Deficit Hyperactivity Disorder

10. What are the best treatments for Attention Deficit Hyperactivity Disorder (ADHD) in teens and adults in both the medical and neuropathic medications?

Question submitted by:
Dr. Dudu Pallie
St. Catherines, Ontario

ADHD or ADD is often an under-recognized and undertreated condition. In part the challenges may lie secondary to the Diagnostic and Statistical Manual of Mental Disorders (DSM) definition that diagnosis is made by seven-years-of-age. It is anticipated that the DSM-V will have the age increased to 13-years-of-age. The patient exists with a triad of symptoms of inattentiveness, impulsivity and hyperactivity. These symptoms exist not only in the pediatric population but the adult population as well. Problems at work, more inattentive at driving, social challenges or substance abuse can be seen as coordinates in the adult world. Often as a clinician, we can get a feel for adult ADHD when they bring their children in secondary to challenges, usually in school, in one of the triad areas of focus.

Treatment options by in large are the same and we must be very aware of transference and counter-transference and be prepared to use appropriate pharmacotherapy and behavioural interventions. In my clinical perspective, adult ADHD is at a point where fibromyalgia was at in the early 1980's.

Long-acting stimulants are first-line therapy according to both the Canadian Attention Deficit Hyperactivity Disorder Resource Alliance and Texas guidelines. Clinically these can be used for adults or children, with caveats being put in place for comorbidities in adults. Stimulants are a logical choice as the ADHD is in part secondary to an imbalance of normal neurotransmitters, especially norepinephrine and dopamine. Concerns with stimulants revolve around abuse potential, but this can be markedly minimized using an Osmotic Release Oral System methylphenidate formulation or a prodrug/amino acid linked D-amphetamine which show a clinically significant reduction in the "likability" index. The likability index is the propensity of a substance to be liked for the potential "high" and could be correlated in direct proportion with abuse lability. These long-acting stimulants only have to be administered once daily to increase adherence, which increases long-term success and enhances functionality. Care should be considered for patients with BP problems as BP and heart rate tend to trend upwards across time with stimulants. The patient's medical comorbidities must be carefully assessed to ensure the right medication is given to the right patient at the right time. Other therapeutic options include atomoxetine or short acting amphetamines, mixed amphetamine salts, and second line options such as imipramine, buproprion or clonidine.

Getting the right medication to the right person at the right time is paramount. Not just in the short-term, but also consider the patient out long-term. There is sufficient evidence to link untreated ADHD to developing bipolar disorder later in life, plus other Axis 1 mental health conditions such as anxiety and depression. As well, there is hypothetical evidence that bipolarity increases the frequency of dementia. As such, functionality in the short and long-term, with effective adherence to the medications and behavioural interventions, cannot be understated.

Answered by:

Prof. Joel Lamoure

11. Schatzki's Ring

What is the significance of a schatzki's ring on barium swallow?

Question submitted by:
Anonymous

Schatzki rings are esophageal mucosal structures at the gastroesophageal junction that are smooth, thin and covered with squamous mucosa above and columnar epithelium below. They are different from muscular rings, which are usually seen in children and located below the squamocolumnar junction and characterized by hypertrophic musculature.

Schatzki rings are often subtle narrowings and are often not appreciated at endoscopy. In fact, a barium swallow and upper GI series is more sensitive at detecting esophageal rings and strictures since these abnormalities will not be seen unless the lower esophagus is widely distended. This may not occur at endoscopy. If an asymptomatic Schatzki ring is found incidentally on a barium study, there is evidence that suggests these rings do not progress. Thus, no treatment is suggested for these patients. Patients who present with dysphagia are usually successfully treated with esophageal dilatation.

Answered by:

Dr. Jerry McGrath

12. *In Vitro* Fertilization Affected by Hydrosalpinx

How does a hydrosalpinx affect fertility and specifically, *in vitro* fertilization (IVF)?

Question submitted by:
Dr. J. Moreau
Barrie, Ontario

A hydrosalpinx is a fluid-filled fallopian tube resulting from infection (chlamydia and gonorrhea), or other inflammatory causes. The presence of a hydrosalpinx negatively affects IVF by up to 30%. Anatomically, it can prevent the union of sperm and egg due to physical blockage. IVF may bypass the blockage but hydrosalpinx fluid can still be extruded into the uterus preventing implantation of the embryo. Furthermore, embryotoxic microorganisms, endotoxins and cytokines are found in hydrosalpingeal fluid. Thus, to improve IVF success rates, the hydrosalpinx should be treated by opening or removing the diseased tube(s).

Answered by:

Dr. Cathy Popadiuk

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