MISSION STATEMENT
The APF’s mission is to provide support to those who have experienced pituitary gland conditions. We promote awareness and disseminate information helpful to the medical community, public, pituitary patients and their families.

FEATUE ARTICLE -
Paediatric Hypopituitarism

Paediatric hypopituitarism is the childhood onset of insufficiency of pituitary hormone secretion. The insufficiency may be partial or complete and may result from pituitary or hypothalamic disease. Pituitary disease or any process that damages the hypothalamus may produce deficiency in pituitary hormones. Symptoms of paediatric hypopituitarism may vary depending on the age of the patient and on the specific hormone deficiencies, which could occur as a single hormone deficiency or in various combinations.

The pituitary gland contains anterior (front) and posterior (back) regions. Most of the hormones that are produced by the pituitary gland are made in the anterior region. The most common hormones produced are: Growth hormone (GH); Adrenocorticotropic hormone (ACTH); Thyroid stimulating hormone (TSH); Luteinizing hormone (LH); Follicle stimulating hormone (FSH); and Prolactin (PRL). Anti-diuretic Hormone or Vasopressin is made in the posterior region.

Cause
Paediatric hypopituitarism has many possible causes resulting from factors that are congenital (present from birth) or that are acquired after birth. Regardless of the cause, all hypopituitarism is characterised by disruptions to the release of one or more pituitary hormones.

Genetic causes of paediatric hypopituitarism are comparatively rare. Research has been conducted that has increased the understanding of the various genetic causes of hypopituitarism that is present from birth.

Congenital causes
Congenital causes of paediatric hypopituitarism can include: perinatal trauma e.g. traumatic delivery, loss of oxygen at birth; absence of or abnormally placed posterior of the pituitary gland; Pallister-Hall syndrome; and interrupted pituitary stalk.

There are genetic disorders that cause hypopituitarism which include: isolated GH deficiency types IA, IB, II and III; multiple pituitary hormone deficiencies (MPHD); isolated gonadotropin deficiency; and Septo-optic dysplasia. There are also defects to the development of the central nervous system that can cause paediatric hypopituitarism.

Acquired causes
Radiation to the brain and hemochromatosis (too much iron in the blood) can result in hypopituitarism. Children who have suffered a traumatic brain injury can also have immediate and delayed effects on their endocrine system.

Infiltrative disorders (build-up of cell substances not normally found in those cells) that could cause hypopituitarism include: Tuberculosis; Sarcoidosis; Lymphocytic Hypophysitis; and Histiocytosis X.

Various types of tumours can also result in hypopituitarism. These include: Craniopharyngioma; Germinoma; Glioma/astrocytoma; and Pituitary adenoma (rare before adulthood).

Epidemiology
Growth hormone deficiency is the most common pituitary hormone deficiency. In 1994 a US study found that...
growth hormone deficiency prevalence of one case in 3480 children. In 2001, a study in Spain estimated the prevalence of hypopituitarism at 4.2 cases per 100,000 adult population. As hypopituitarism has both acquired and congenital causes, it can occur in neonates, infants, children, adolescents and adults.

**Prognosis**
The overall prognosis for paediatric hypopituitarism, with the appropriate treatment, is very good. Long term complications can include osteoporosis, increased cardiovascular disease, short stature, and infertility.

Recognition of pituitary hormone insufficiency and appropriate hormone replacement therapy is vital for avoidance of complications and, in some cases, death. Single or multiple deficiencies of pituitary hormones can result in significant chronic conditions, including:
- **Adrenal crisis** (due to lack of cortisol hormone) - can occur when the patient is under considerable stress, from ACTH deficiency, symptoms include low blood pressure, low blood sugar or hypoglycaemia, severe shock and death.
- **Short stature** - could have considerable psychosocial consequences.
- **Hypoglycaemia** - could cause convulsions, persistent and severe hypoglycaemia can cause permanent central nervous system injury.
- **Osteoporosis** - increased risk of fractures.
- **Hypogonadism and impaired fertility** - from gonadotropin deficiency.

Growth hormone deficiency is understood to be an important contributing factor to the disability and death associated with hypopituitarism. Causes of disability and death are often related to many factors and related to the specific cause of paediatric hypopituitarism, also taking into account the degree of pituitary hormone deficiency.

**Clinical presentation**
The symptoms of paediatric hypopituitarism will vary depending on the patient’s age, the cause, and the hormones that are deficient.

**Neonates**
Most babies with hypopituitarism will have normal to high birth weights and lengths. Most will also not have any history of slow growth during their development in the uterus. Babies often have presentations of breech birth, however the reasons for this are still unclear. Micro-penis in males can result from a deficiency in gonadotropin or growth hormone.

Babies with hypopituitarism are at a higher risk of hypoglycaemia. Symptoms include jitteriness, convulsions, lethargy, pale skin, bluish discolouration of the skin from a lack of oxygen in the blood, and the temporary stopping of breathing.

Jaundice can also occur when a baby has hypopituitarism. If a full term infant has unexplained temporary stopping of breathing, low blood pressure or unstable temperature, then these are possible symptoms of hypopituitarism. Electrolyte disturbances can also be a symptom.

**Older infants and children**
Common symptoms of hypopituitarism in older infants and children include failure to grow, delayed or early puberty, and diabetes insipidus due to ADH deficiency.

Lack of growth is the most common symptom of hypopituitarism, and can also be associated with a delay in tooth development. Hypoglycaemia in neonates or young infants can also be another symptom. Patients with acquired or mild forms of gonadotropin deficiency, who do not show micro-penis in infancy, may have absent or delayed puberty, including in girls primary or secondary amenorrhoea or lack of menstrual periods.

Diabetes insipidus can be difficult to diagnose in infancy due to nonspecific symptoms, but is usually associated with poor weight gain or failure to thrive and electrolyte disturbance. Symptoms of diabetes insipidus, such as large volumes of urine and abnormal thirst, are easier to diagnose in older children.

Depending on the cause of the hypopituitarism, associated problems in the infant or child include visual symptoms, neurodevelopmental delays, seizure disorders and some congenital malformation syndromes.

**Diagnosis**
Blood tests are necessary in the diagnosis of paediatric hypopituitarism. It is recommended that any patient with hypopituitarism has a magnetic resonance imaging (MRI) study to exclude the possibility of a brain tumour. For hypopituitarism, the preferred imaging study is a brain MRI with specific cuts of the pituitary gland.

Testing for growth hormone deficiency is usually done using insulin-like growth factor-1 (IGF-1) and insulin-like growth factor binding protein 3 (IGFBP-3), except in the case of brain tumours. There is no diagnostic value to random measurements of GH levels except in early infancy. If growth hormone deficiency is suspected, the further testing of GH secretion is ordered under the direction of a paediatric endocrinologist.

To exclude the possibility of a CRH-ACTH-cortisol axis deficiency, measuring a patient’s morning serum cortisol levels can be useful. For patients with hypoglycaemia, a blood sample showing low serum glucose, and measuring cortisol levels and GH, can be diagnostic of hypopituitarism. To determine hypothyroidism, diagnosis is based on low free thyroxine (FT4) and low serum TSH levels.

The approach to testing the pituitary-gonadal axis is based on patient age. To test young infants, random measurements of testosterone or oestradiol levels and of FSH and LH are adequate, but is best done in the early neonatal period up to 3 months of age. For patients aged in later infancy to 4 years GnRH testing is an option. There are no reliable tests for a pre-pubertal child so testing is...
Treatment

The primary treatment for hypopituitarism is the appropriate hormone replacement. The medication choice is determined by the presence of one or more hormone deficiency. It is also necessary to stress dose using corticosteroid replacement when suitable. Surgery can be used in cases of tumour associated hypopituitarism, with the location of the tumour directing the choice of surgical procedure.

Special dietary considerations are necessary in the treatment of some children with hypopituitarism. If the child also has diabetes insipidus, their water and fluid intake may require close monitoring. Children with hypopituitarism and hypothalamic damage can be at risk of rapid weight gain leading to obesity. These children need their daily intake of food closely monitored.

Patients with hypopituitarism need their growth and development routinely monitored. If the patient is on growth hormone therapy then they need to be monitored for any adverse reactions. Patients with hypopituitarism should have their thyroid functions monitored. Patients with hypopituitarism, which also includes adrenal insufficiency, should wear some form of medical alert identification, to inform emergency personal to the need for corticosteroid replacement when suitable. Surgery can be used in cases of tumour associated hypopituitarism, with the location of the tumour directing the choice of surgical procedure.

Medication used for the treatment of hypopituitarism simply replaces the deficient hormone(s). Dosing is determined by what is the normal, healthy levels that a hormone should be functioning at. If administered correctly, adverse effects are not common. Consistent and precise compliance of prescribed medications is essential to avoid too much or too little hormone.

References


Reviewed for APF by Dr Gary Leong, Lady Cilento Children’s Hospital, Brisbane.

Don’t Forget! New contact details for state coordinators

1300 307 866 at the cost of a local call if you live in the same state as your coordinator. If you wish to contact APF personnel outside of your state, or discuss APF business or contact support, please ring the national phone number of 1300 331 807.

Patient Story: Simon

My story has been a lifelong one and while it has come with a sense of isolation, the ongoing mission has been constantly on the path to catch-up to the pack!

I was born with congenital panhypopituitarism (“panhypopit”) with no previous family history and not diagnosed until three years of age. My parents found themselves with a son who was quite lethargic and short in stature, however with no diagnosed reason behind it. After a number of tests at the RCH in Melbourne, panhypopit was identified as my condition, and commenced treatment of growth hormone through delivery from an intramuscular injection. I’ll be forever grateful to the paediatric endocrinologists of the RCH, Melbourne for their talent, dedication and total patient care for what I have today.

Commencing the growth hormone program was met with excitement, trepidation and the start of many painful nights for both patient and parent! The growth hormone program was suspended for 12 months when treatment reverted to Somatinorm, I felt I lost some momentum. I spent much time hovering below the bottom line of the height graph; however the results eventually came in reaching my genetic-determined height at the age of seventeen, although it took a lot of perseverance and courage. Emotionally and physically though I always felt a few years behind others at school and with a condition not well known and experiences that I felt couldn’t be shared, I spent a child hood feeling particularly isolated.

At the end of Year 10, I considered whether it would be worthwhile continuing at school or to start finding work. Being somewhat of a “late bloomer” with school work, the penny didn’t drop until a couple of years after Year 12, however while at University I studied particularly hard and have since gone on to achieve CPA status which I feel particularly proud of. I had an overwhelming drive to catch-up for the years I felt I wasn’t ready for back in High School.

After settling into a professional career, I met my now wife and we commenced preparations for starting a family, unsure whether we would achieve an outcome. I had three goals in life (i) to gain a professional qualification; (ii) to be a husband; and (iii) to be a “daddy”. However with no guarantees of being able to have children, I felt there would have been a void unable to fill.

I commenced regular self-injected intramuscular and subcutaneous injections in January 2008. As much as I didn’t enjoy reverting back to a regular injecting program, I felt I wanted to experience the sacrifice of what was a painful experience in order to achieve life goal # 3. And in October, 2009, God blessed my wife and I with the arrival of our first daughter, then again with our second daughter in April 2012.

When our second daughter was born (Georgie), she was taken to ICU hours after birth with episodes of...
hypoglycaemia and hypothermia. Georgie’s blood-sugar results continued to return low numbers in her first couple of days, and with my condition being a possible link, we committed to having cortisol and growth hormone tests conducted which as a parent is particularly harrowing. Results confirmed her as diagnosed with the condition congenital panhypopituitarism. I felt relieved we had an answer and an action plan for her immediate health, coupled with a sense of personal guilt and sadness she had my condition. However, in situations such as these, rational thoughts of “what medication dosage to give” and “what blood-sugar results are within an acceptable level” took precedence and I’ve come to realise we simply need to work with this as a family unit – put simply, whatever life throws your way.

Since becoming a parent of a child with panhypopit, life takes a very different slant and both I and Georgie had an MRI conducted which presented that we were both born without a pituitary gland. Thankfully Georgie is now a very healthy girl and her development continues to evolve.

Being born during these times and in this country I feel we’re both very fortunate to be (a.) diagnosed and (b.) treated, and continue to love a normal life.

I started my life feeling isolated, however with the love and support of family and friends, and now with Georgie, together we’re stronger.
members, and very importantly a new little friendship was formed by 2 children with pituitary disorders.

Next get-together? Possibly around September when it starts to warm up - will advise you when confirmed.

Sue

**NSW**

**Social News:** Thanks to everyone who turned up to our latest get together. It started with a look around the discovery museum at the Rocks, Sydney, which I think the kids liked especially. The treasure hunt, which was to learn about the history of the rocks area in Sydney, added to the entertainment.

Afterwards we met you at the Observer Hotel for lunch. I don’t know about everyone else but my lamb burger was beautiful.

It was good to see familiar faces and some new ones as well. Hope to see you at our next get together or at a seminar.

Daniel

**WA**

**Social News:** We will be having information booths at the hospitals in Perth the week of July 27th to 31st to celebrate “Pituitary Awareness Week” Thanks to those who have offered to attend with me – I look forward to seeing you all.

**Seminar News:** Saturday 1st August – a combined adult and paediatric education day at the Auditorium at Sir Charles Gairdner Hospital.

**For the Adult:** Doctors at Sir Charles Gairdner Hospital have kindly offered to make presentations to patients and members on Saturday 1st August at the Mary Lockett Lecture Theatre.

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<thead>
<tr>
<th>Time</th>
<th>Session</th>
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<tr>
<td>8.45 - 9.00am</td>
<td>Registration and a cuppa</td>
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<td>9.00 - 9.10am</td>
<td>Greeting and Introduction : Sue Kozi and Peter Marsh, APF</td>
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<td>9.10 - 9.50am</td>
<td>Role of the adrenal gland, hydrocortisone and sick day management: : Dr. David Henley, Endocrinologist, Sir Charles Gairdner Hospital</td>
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<td>9.50 - 10.30am</td>
<td>Living with and managing Prolactinoma : Dr. Joey Kaye, Endocrinologist, Sir Charles Gairdner Hospital</td>
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Vic

**Social News:** Jack Kagan certainly picked a great spot on Saturday 28th March at the Boat Builders Yard Café, South Wharf. It was a typical beautiful Melbourne autumn day. Jack reports….

10 people attended. The aim of the discussion was to address the fact that “we are not alone”. We mixed up the discussion by changing where people sat, so that everyone could talk to everyone else. As always it was apparent that people needed to talk about their unique experiences. Although most people at the table did not know each other, by the end of the afternoon everyone had created a new friend. The value of these discussions cannot be underestimated.
To know that there is someone to turn to, to discuss what you believe is unique, is a cathartic experience.

Victoria does not have a state co-ordinator and is looking for a person to assist with planning the various functions that we regularly have. Is anyone prepared to put up their hand? Visit our website to find out more www.pituitary.asn.au

**Seminar News:** Saturday 8th August we will be hosting a combined adult and parent education day in conjunction with Bendigo Health at the Monash School of Rural Health.

**For the Adult:** Information on thyroid, adrenal and well-being will be the focus in the morning. Many people know that the pituitary regulates other endocrine glands, but not exactly what those endocrine glands mean for good health. The adult session will conclude with a workshop on everyday strategies for coping which will be exciting.

8.45 - 9.15am Registration and a cuppa
9.15 - 9.30am Greeting and Introduction : Sue Kozij and Peter Marsh, APF
9.30 - 10.20am The role of the thyroid gland
What is the difference between primary and secondary hypothyroidism?
The role of the adrenal gland
What is the difference between Pituitary Adrenal deficiencies, Addison's Disease and Addisonian crisis
Sick day management and hydrocortisone
Brief update on new formulations of hydrocortisone being researched :
Dr. Mark Savage, Endocrinologist, Bendigo Health
Dr. Carmela Caputo, Endocrinologist, St. Vincent's Hospital, Melbourne
10.20 - 10.45am Morning Tea
10.45 - 12.00pm Coming to terms with a "new normal me"
: Rachel Kane, Psychologist
12.00 - 12.30pm Discussion
12.30 - 1.30pm Light Lunch

**For the parents of children with pituitary disorders:**
Join us to discuss various issues around problems with their pituitary gland; why the child needs to take the drugs that are prescribed; what to do on sick days and why; how and when to transition responsibility for medicine from the parent to the adolescent; what do extended family, friends' parents, kinder and school need to know.

1.00 - 1.30pm Registration
1.30 - 1.45pm Greeting and Introduction :
Sue Kozij, Simon White APF and Dr Andy Lovett, Clinical Director of Paediatrics, Bendigo Health
1.45 - 3.15pm :
A/Professor Christine Rodda, Paediatric Endocrinologist, Sunshine and Ballarat Base Hospitals:
Dr Andy Lovett, Paediatrician, Bendigo Health
3.15 - 3.45pm Questions
3.45 - 4.30pm Afternoon Tea

**GPCE:**
Sue, Daniel and Beth have just completed the "manning" of an information booth at the Sydney General Practitioner Conference and Exhibition at The Dome, Sydney Olympic Park from Friday 22nd to Sunday 24th May, which also included the PNCE - Practice Nurse Clinical Education - running alongside the GPCE, affording us the opportunity to educate and inform specialist nurses as well. Our theme for this event was "Could it be the Pituitary? - The Master Gland", aiming to inform and educate GP's about early detection of pituitary conditions, and how many pituitary symptoms, at times, mimic other problems.

The enormous effort to organise this educational event for GP's was well worth it!! Although "walk by" traffic was not as high as we hoped, the workshops were a huge success!! All three workshops were extremely popular and filled to capacity!!

On speaking with GPs we list a couple of observations:
- GPs think that pituitary tests are expensive to run
- Because some endocrine and other symptoms are common, they run the more common tests first eg. Thyroid function as a singular test, rather than a full pituitary workup
- The likely hood of coming across a pituitary disorder is very low, so they don’t think it a priority to run the tests.

Thank you so very much to our wonderful supporters who presented for us – how could we do this without you!!!
We were in a position to offer a free delegate’s registration to the GPCE in Sydney.

Dr Charlotte Hespe from Glebe Family Medical Practice attended and these were her comments.

“From my perspective having a stall there to draw attention to pituitary disorders was a really great idea.

The workshop was perfect for GP’s- interesting cases that showed the diagnostic dilemmas we face with patients presenting with non-specific symptoms and the difficulties we might have in interpreting hormone levels.

The case studies gave us a good framework to assist in future diagnosis and appropriate management / referrals.”

Thanks to our sponsors which funded our attendance at this event

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**Membership**

Most members will have their financial membership expire by the end of the month. Can we please encourage you to re-join so that you can support us in the work we do? It can now all be done on-line. No more filling out the form each year – you can simply go to your profile and update it, along with selecting your preference with the contact support register. We subscribe to a safe payment gateway run by Commonwealth Bank. If you have not registered on the website yet – please select the log in, then register, complete the information and it should take you to a welcome page – there it will ask you to link your profile within the database. If you have difficulties please email membership@pituitary.asn.au

Additionally it is the end of the financial year. If you can spare a small tax deductible donation to APF that would be fantastic – or do you have any personal or corporate contacts who are looking to support a charity? http://pituitary.asn.au/GetInvolved/CorporateSupport.aspx

**Fundraising**

Receive over $20,000 worth of valuable offers from the best local restaurants, your favourite cafés, hotels and resorts, travel, attractions, shopping and more.

1. Enjoy hundreds of 2-for-1 and up to 50% off offers from the best restaurants and cafés in your area.
2. Discover the exclusive offers from Virgin Australia and Emirates Airlines for amazing value when you fly.
3. Treat yourself to a shopping spree with great offers.
4. Be ‘entertained’ all year long with valuable cinema and theme park offers.
5. Feel the reward of your purchase contributing to community fundraising!

Purchase your Entertainment™ Book from Australian Pituitary Foundation again to support our fundraising efforts this year.

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**Teleconferencing Support**

A great session was done on an evening in March. Attendees held many discussions on non-hormone/ functioning tumours, including various treatment options offered to them on re-occurrence. It is wonderful to see that a couple of these people have continued to stay in contact privately.

Further sessions will be held on separate evenings for: 1) parents of children (general); 2) Specific topics regarding puberty, adolescence and boys. 3) Prolactinoma.

Please see the events area of our website for further details.

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**New look for our website**

Volunteer Beth has been extremely busy redesigning our website. Please go to it to have a look – WOW. www.pituitary.asn.au More enhancements will be made to the fact sheet area over coming months. Please encourage your GP to look at the Health Professionals tab, they will need to register to gain access.
Recently Sue visited New Zealand for a holiday and took the opportunity to catch up with Catherine Chan, Chairperson, from The New Zealand Acromegaly Society.

The two have met only once before, but certainly have shared ideas and resources over many years.

We are so fortunate to have a professional Copy Writer contact us with the offer to write resources for us. Firstly, we intend to increase paediatric resources and just recently conducted a meeting in Melbourne of interested Mums and Dads of children with congenital hypopituitarism.