CENTRAL PRECOCIOUS PUBERTY: Challenges and opportunities for nurses

In a webinar held on 28 November 2020 paediatric endocrinology nurses Kate Davies and Lee Martin from London, UK shared their knowledge and experiences in managing and caring for patients with central precocious puberty. With an agenda specifically crafted by nurses for nurses, the IPSEN-sponsored webinar also provided participants a peek into a nurse’s journey to specialising in paediatric endocrinology.

All about Normal Puberty

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Puberty is the development of secondary sexual characteristics in order to attain reproductive capacity. It is characterised by androgen-dependent changes in the body (adrenarche), breast development in females (thelarche), gonadal changes in response to pituitary gonadotropins (gonadarche), pubic hair growth (pubarche), onset of menstruation in females (menarche) and the development of sperm in the male testes (spermatogenesis).

Normal puberty starts after the age of 8 years in girls (average is 10 years) and after the age of 9 years in boys (average is 12 years). Genes that have been studied in relation to central precocious puberty (CPP) include MKRN3, DLK1, and KISS1R, while IGFS10 has been associated with delayed puberty.1 Obesity also affects the timing of puberty; if a child is obese, puberty may start earlier; conversely, if a child is too thin, onset of puberty may be delayed. Emotional and behavioural difficulties, as well as chronic disease, may also have an impact on the onset of puberty. The onset of puberty in children who have undergone craniospinal, gonadal, and abdominal irradiation for cancer may be affected. Environmental exposure to phytooestrogens, dichlorodiphenyltrichloroethane (DDT) and bisphenol A (BPA) have also been purported to have effects on puberty.2

Recent studies have shown that over the last 50 years, the onset of menarche has been occurring increasingly earlier (Figure 1).3 The age at thelarche has also shown a downward trend of 0.24 years per decade from 1997-2013.4 In Korea, the incidence of CPP also appear to be rising (Figure 2).5 While the exact cause has not been established, factors that have been identified as possible contributors to the rise in incidence are genetics, environmental factors, and increase in obesity.

Early puberty and management of central precocious puberty (CPP)

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Precocious puberty is when a child’s body begins to change into that of an adult too soon. It is considered when puberty begins before age 8 in girls (breast development at stage B2) and before age 9 in boys (testicular size 4ml bilaterally).

Central precocious puberty is defined as pubertal development due to early activation of the hypothalamic-pituitary-gonadal (HPG) axis (Figure 3, left). In contrast, pseudo-precocious puberty occurs when pubertal development is caused by sex steroid secretion without the activation of the HPG axis (Figure 3, right).

Figure 1. Declining age at menarche in Europe – from approximately 17 years in the early 19th century to approximately 13 years by the mid-20th century.

Figure 2. Annual incidence of central precocious puberty in Korean children from 2008 to 2014.

Figure 3. Hormonal changes driving CPP (left) and pseudo-precocious puberty (right).
Management of CPP – Case Vignettes

CASE 1: A 2.2-year-old female presented with breast development at 10 months of age and pubic hair development at 12 months of age. Pubertal assessment revealed Tanner Stage 2, and bone age on x-ray was 6.6 years (Figure 4). Biochemical investigations revealed elevated levels of LH (1.1iu/L), FSH (5.0iu/L) and oestradiol (87pmol/L). A gonadotropin releasing hormone (GnRH) stimulation test showed elevated levels of both LH and FSH. A diagnosis of CPP was made and treatment with GnRH analogues was started. This resulted in decreased release of LH and FSH and consequently, decreased levels of oestrogen.

CASE 2: A 4-year-old female presented with vaginal bleeding for 3 days associated with lower abdominal pain. Physical examination revealed a suprapubic mass. Tanner staging was B3A2P2 (Figure 5) and the vagina appeared oestrogenised. Bone age was 6.5 years and biochemical investigations revealed a highly elevated oestradiol level (924pmol/L). The GnRH stimulation test was unremarkable. This was diagnosed as pseudo-precocious puberty. Abdominopelvic MRI scan revealed a large abdominal mass for which the patient underwent laparotomy and right oophorectomy due to the large cystic mass replacing the right ovary. After the removal of the mass, oestrogen levels became normal and follow-up monitoring revealed normal pubertal development thereafter.

My journey to becoming a CPP nurse

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Decades ago, there was a nursing stereotype that nurses are doctors’ ‘handmaidens’; however, in recent years, nursing has become a more autonomous profession. Nurses manage their own patient caseloads in nurse-led clinics, with some nurses running their own thyroid/ adrenal/late effects clinics. Nurse-led clinics reduce patients’ waiting times, facilitate prompt prescription of growth hormone, and allow consultants to focus on more complex cases. Nurse-led clinics also allow nurses to build stronger relationships with patients and their families, which enhances patient satisfaction.

There are many opportunities for paediatric nurses to gain advanced skills in CPP patient care, including physical and bone age assessment and independent prescribing. “I began as a children’s nurse in different hospitals until I took a special interest in paediatric endocrinology while working at the Great Ormond Street Hospital in London. I pursued further education and training and am now currently registered with the Nursing & Midwifery Council as a children’s nurse, teacher, and nurse independent or supplementary prescriber. I have also now been involved in the development of several publications on CPP,” Ms Davies shared.

A paediatric endocrine nurse takes on many roles: patient education and support for the patients and their family, hands-on nursing care, facilitation of dynamic function tests, liaising with different parties involved in patient care, teaching and research. Apart from being a clinical expert, CNS are also patient advocates and agents of change. The success of clinical nurse specialists as change agents depends on their ability to develop their own support system and find job satisfaction. A high level of motivation is key, as well as the ability to negotiate and push for change. “Change may come gradually, but it is well worth it because ultimately, it will benefit patient care – the heart of what we do,” Ms Davies concluded.

For more information about CPP, visit: sci.ipsen.asia

Q & A

Does menarche signal the start of puberty?
No, to the contrary, menarche signifies the end of puberty. It is the last stage of puberty for females.

What advice would you give to parents who notice early pubertal changes in their children?
Any signs of early puberty need to be investigated. The challenge is getting parents to recognise that it is a problem that needs to be addressed. If families have any doubts, they should see a professional.

How can we do more for our CPP patients?
Paediatric endocrine nurses have come a long way over the last several decades. Their capabilities can be enhanced through further education. While independent nurse endocrine services offer advantages, it is important to do things in conjunction with consultants. Multidisciplinary meetings are essential in managing patients most appropriately.