

Hong Kong Journal of Paediatrics

香港兒科醫學雜誌 (New Series)

An Official Publication of
Hong Kong College of Paediatricians &
Hong Kong Paediatric Society
c/o Hong Kong College of Paediatricians, Room 801,
Hong Kong Academy of Medicine Jockey Club Building,
99 Wong Chuk Hang Road, Aberdeen, Hong Kong.

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Hong Kong Journal of Paediatrics is published by
Medcom Ltd, Flat E8, 10/F, Ka Ming Court,
688-690 Castle Peak Road, Kowloon, Hong Kong
SAR. Tel: (852) 2578 3833,
Email: mcl@medcom.com.hk

Indexed in EMBASE/Excerpta Medica, Science
Citation Index Expanded (SCIE) and Scopus

Website: www.hkjpaed.org

ISSN 2309-5393 (online)

ISSN 1013-9923 (print)

Editorial

Managing Complicated Diseases with Basic Approach

With the rapid advancement of science in modern era, the management of complicated diseases is becoming more sophisticated and expensive. Such as the use of gene therapy for spinal muscular atrophy, targeted therapy for cancers, biological blocking agents for refractory autoimmune diseases, etc. All of these are exciting achievement but not all clinical problems can be resolved with such approach, and we should not forget the basic. The original articles in this issue are supporting this concept.

Necrotising enterocolitis (NEC) is a challenging condition in neonates, especially for the premature neonates. Early detection may help to prevent extensive damage to the bowel. In recent years, many bio- or genetic markers have been advocated but few has reached the actual clinical application threshold. Faecal calprotectin has been advocated as a marker for inflammatory bowel disease and other bowel inflammatory condition such as NEC, but again it is non-specific. Calprotectin is a calcium and zinc binding protein found within the neutrophils and the elevated calprotectin level in the stool simply reflects neutrophil migration to the bowel tissue during an inflammatory process. Therefore, one must be aware of other confounding variables. Cekovic et al evaluated the meconial calprotectin of both normal or premature neonates and found that its level can be affected by gestational age, asphyxia, feeding intolerance, hyperbilirubinemia, and metabolic disturbances. However, tests can be either sensitive or specific. Since faecal sample is non-invasive and elevated calprotectin level in the gut suggests bowel inflammation, it can still be a reasonable reference. This simple test may be helpful not only for diagnosis but also monitoring the disease process. However, the threshold of significant level remains to be determined by future investigations.

Biliary atresia is a life-threatening condition if not manage properly. Due to the impaired liver function caused by the obstructed biliary outflow, it may affect the enteral absorption. Therefore, many of the affected infants are given total parenteral nutrition instead of enteral feeding. However, such practice induces secondary problem such as enteral cells atrophy and malabsorption in long term. Wu et al performed a randomised study and showed that early enteral nutrition can improve the nutritional status and growth of children following the Kasai procedure. In addition, it also facilitates the gastrointestinal flora balance, reduces the number of postoperative cholangitis episodes, and improves life span of the diseased liver. These findings are strong support of reintroducing the enteral feeding early for children with biliary atresia after Kasai operation. The belief of avoiding overloading the "diseased" liver after Kasai operation is probably a myth and we should base our practice on scientific evidence.

Though rare locally, cystic fibrosis is a relatively common chronic

disease for Caucasian population. It affects multiple organs with complications including recurrent respiratory infection, pancreatic insufficiency, etc. For the lung condition, patients often require long term antibiotics coverage and respiratory care such as chest physiotherapy. Consequently, patients and their parents are often under a tremendous amount of stress leading to significant anxiety and depression. The current management is mainly focusing on the treatment of the patients and may miss the other important aspects of life including the wellbeing of the patients and their care providers. Kalyoncu et al provided a 12-session online breathing and relaxation training program to 68 patients and their families. The training was provided by a psychologist. The duration of each session was one hour per week and participants were advised to practice daily after the training. Then multiple questionnaires were used to assess the potential changes. It was shown that breathing and relaxation exercises may help to alleviate depression and anxiety, leading to a better life satisfaction, optimal sleep pattern for patients and their parents. Although this is a preliminary study with small sample size, breathing exercise and relaxation technique is a simple and cheap method to enhance wellbeing, patients and their parents are encouraged to learn more about this technique and practice it on a regular basis.

Hearing impairment may have a high impact on children especially for their learning and development of interpersonal relationship. Hearing loss can be caused by either environmental and/or genetic causes. Genetic causes can be syndromic or non-syndromic related. Non-syndromic genetic causes are more common (>70%) and up to 80% of them are due to mutation in the *GJB2* gene

which is inherited in an autosomal recessive manner. Other genetic aetiologies include autosomal dominant, X-linked and mitochondrial variants. It is well documented that aminoglycoside such as amikacin, gentamycin or neomycin can lead to hearing impairment in a selected group of patients and the old belief is that they are "environmentally" related. As our knowledge improved, it becomes known that such predisposition is in fact genetic related. Mitochondrial *m.1555A>G* mutation is associated with maternally-inherited aminoglycoside-induced hearing loss and non-syndromic deafness. Leung et al, reported a cohort of 14 local family with this genetic defect. It is fortunate that none of this cohort have previous exposure to aminoglycoside. Since such genetic defect is known and test is readily available, should we check for such defect for our patients before we consider using aminoglycoside? Similar precaution has been applied to the use of carbamazepine (HLA-B*1502 allele for Steven-Johnson syndrome), allopurinol (HLA*58.01 for Steven-Johnson syndrome), and azathioprine (TPMT or NUDT-15 deficiency for myelosuppression) nowadays in the local Hospital Authority system.

As illustrated by these studies, basic diagnostic and therapeutic approach can still be very helpful for our clinical management, even for complicated and chronic illnesses. While we embrace the advanced technologies, one has to remember the basic and they can still be very useful for our daily medical practice.

GCF CHAN
Chief Editor