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Editorial

Prognostic Indicators of Diseases

In this issue, several studies applied clinical observations to predict the possible outcome of their respective patients. It has been reported that thyroid agenesis is associated with an increase incidence of structural abnormalities in other non-thyroid organs. But in the literature, such association can range from 8% to more than 50% in different ethnic populations. Whether there is any genuine racial predisposition remains to be answered for most studies are not population based. Current evidence also suggests that some commonly shared developmental genetic pathways are likely to be involved in such association. Therefore, the prognostic significance of thyroid dysgenesis with or without extra-thyroid involvement may be related to the underlying genetic anomalies. Additional genetic investigations may help to answer this question.

Another study tried to define the severity and associated cardiovascular abnormalities of different types of vascular ring based on the anatomical parameters. It was found that pulmonary artery sling (PAS) is associated with higher incidence of stridor and dyspnoea as compared to patients with right aortic arch. The diagnosis can be best assessed with CT contrast angiography or MRI. It was also found that PAS is associated with a higher frequency of additional cardiovascular anomalies. One of the key learning points of this study is the possible misdiagnosis of either "asthma" or "laryngitis" of this group of patients leading to delayed diagnosis. Chronic cough is the most common presenting symptoms and was found in all patients with double aortic arch. Aware of this clinical association will help to minimise unnecessary delay.

Burst suppression pattern as identified by EEG in neonates have been considered an ominous sign. However, there are isolated situations that the prognosis of this anomaly can be less severe. The underlying pathological conditions are likely affecting the prognosis. Shen et al. reviewed their patients' cohort and found that neonates with hypoxic ischaemic encephalopathy (HIE) have relatively better outcome as compared to those with Ohtahara syndrome (OS) or early myoclonic encephalopathy (EME). In fact, both OS and EME should not be concluded as the final diagnosis for both of them have heterogeneous etiologies. A wide variety of genetic or structural anomalies have been identified with OS and another spectrum of metabolic conditions can be associated with EME. While HIE is a non-progressive form of injury, most of the OS and EME are progressive in nature if the underlying etiologies cannot be rectified. It is known that some of the underlying conditions can be readily treated now such as pyridoxine related disorders leading to EME. Proper diagnosis with early appropriate management will help to alter the outcome drastically for some of

these children. With more understanding of the underlying genetic mechanisms, we foresee more targeted therapy becoming available in the near future for these disorders. One of the breakthroughs in recent years is the use of mTOR inhibitor in children with tuberous sclerosis, they often present with infantile spasm in early childhood and have refractory seizure associated with progressive deterioration in their cognitive function during their later life. Recently, the early use of mTOR inhibitor has the potential in controlling the progression of cortical tuber with subsequent better seizure control and improvement in cognitive function. This type of approach may guide us in changing the prognosis of some of the children with OS and EME in the future.

For patient with septic shock requiring ICU care, early use of potent (i.e. epinephrine) or multiple inotropes and higher initial / increase fluctuation in monocytes count are all indicators of poor survival as mentioned by Delgado et al. The early use of potent and multiple inotropes may not be a surprise indicator for that reflects the severity of shock and these patients were probably approaching the end of the "shock spiral". But the monocytes count seems relatively controversial for elevation of leucocytes and platelets are well known indicators of stress. In severe sepsis, it is often associated with disseminated intravascular coagulopathy and we expect neutropenia and thrombocytopenia as a consequence. This study highlighted the importance of looking at monocytes for it may be related to the severity of

the subsequent cytokine storms. Since antigen presenting cells such as macrophages and dendritic cells are derived from monocytes, elevated monocytes count implies that there are more active adaptive immune responses ongoing with increase cytokines production. Whether this will be a useful prognostic indicator remains to be verified. Means to suppress cytokine storm involves the use of immunosuppressants and even chemotherapy such as etoposide. Such approach has been tested in the setting of acquired hemophagocytic lymphohistiocytosis. This will be a paradigm shift in the management of septic shock if proven to be a major prognostic determinant.

Therefore, in clinical medicine, we have been relying on known clinical parameters to predict the outcome of our patients and to guide us on further management. Many of the underlying mechanisms of these clinical predictors can now be explained by either genetic or immunological defined abnormalities. Understanding of these mechanisms will help us to design more targeted and effective management of our patients. With the genetic and immunological tests getting more readily available at an affordable cost, we expect a new chapter of medical care will be ahead of us in the near future.

GCF Chan
Chief Editor

Erratum

In the last Editorial, the first line "chief compliant." should be corrected for "chief complaint". Apologise for the typo.