Proliferation of syndromes and acronyms in paediatric critical care: are we more or less confused?

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As with many disciplines in medicine, syndromes, abbreviations, and acronyms have been coined to aid diagnosis and prognostication in paediatric intensive or critical care medicine (CCM). Although these acronyms are commonly used, they remain controversial. In clinical practice, CCM syndromes are clinical patterns resulting from the interaction between insult and host response. They are closely correlated to organ system failures and are common presentations of diverse aetiologic factors. Therefore, it is important to search for underlying aetiologies and subtypes. In research, the role of acronyms and syndromes is limited in epidemiological studies and clinical trials enrolling patients meeting syndrome criteria, unless underlying subtypes and aetiologies are considered. As a result, study outcomes are often negative. Hence, we advocate that both syndrome codes and underlying aetiologic diagnoses be provided in International Classification of Diseases (ICD) coding to record prognostic and therapeutic significance (Table). Last, non-standardised acronyms contribute to confusion and should be used sparingly. Herein, we review some examples of syndromes and acronyms in use in CCM settings and consider whether their use is justified.

Acute respiratory distress syndrome (ARDS) is an important cause for CCM admission.1,2 Ashbaugh et al3 first described ARDS as an adult-type respiratory distress for a group of patients with progressive respiratory failure, refractory hypoxaemia, decreased functional residual capacity and lung compliance, and diffuse infiltration on chest radiography. In 1994, the American-European Consensus Conference introduced ARDS as a disease with acute-onset hypoxaemia, PaO2/FiO2 ratio ≤200, bilateral infiltrates on chest radiographs, and the absence of left atrial hypertension.4 The international consensus criteria for ARDS were updated in 2012 and are known as the Berlin definition.5 However, ARDS is not a specific disease entity; it is a clinical syndrome which may be triggered by various pathologies such as trauma, pneumonia, and sepsis. As with many other syndromes, the term essentially describes non-cardiogenic pulmonary oedema of various aetiologies.

Acute respiratory distress syndrome also occurs in children, and is no longer restricted to adults.6 In 2015, the Paediatric Acute Lung Injury Consensus Conference proposed a definition for paediatric ARDS.7 This definition identified more patients with ARDS than did the Berlin criteria, but there were no differences in clinical outcomes.8 However, the Berlin definition offers no room for stratifying and identifying true ARDS patients because there is no re-evaluation of hypoxaemia under standard ventilator setting in a specific time period.9

The current (2015) ICD code for ARDS is

### TABLE. Selected acronyms used in critical care medicine setting and their ICD codes

<table>
<thead>
<tr>
<th>Acronym</th>
<th>ICD-9-CM code</th>
<th>ICD-10-CM code</th>
</tr>
</thead>
<tbody>
<tr>
<td>ARDS</td>
<td>518.82</td>
<td>J80</td>
</tr>
<tr>
<td>COVID-19</td>
<td>U07.1</td>
<td></td>
</tr>
<tr>
<td>GVHD</td>
<td>279.5</td>
<td>D89.81</td>
</tr>
<tr>
<td>MERS MODS</td>
<td>995.2</td>
<td>R65.2</td>
</tr>
<tr>
<td>Pneumonia due to COVID-19</td>
<td>J12.89</td>
<td></td>
</tr>
<tr>
<td>PRES</td>
<td>67.83</td>
<td></td>
</tr>
<tr>
<td>SARS</td>
<td>079.82</td>
<td>B97.21</td>
</tr>
<tr>
<td>Severe sepsis</td>
<td>R65.2</td>
<td></td>
</tr>
<tr>
<td>SIRS</td>
<td>R65.1</td>
<td></td>
</tr>
<tr>
<td>SSSS</td>
<td>695.81</td>
<td>L00</td>
</tr>
<tr>
<td>TLS</td>
<td>277.88</td>
<td>E88.3</td>
</tr>
<tr>
<td>TMA</td>
<td>446.6</td>
<td>M31.1</td>
</tr>
<tr>
<td>TSS</td>
<td>040.82</td>
<td>A48.3</td>
</tr>
<tr>
<td>VOD</td>
<td>573.8</td>
<td>K76.5</td>
</tr>
</tbody>
</table>

Abbreviations: ARDS = acute respiratory distress syndrome; COVID-19 = coronavirus disease 2019; GVHD = graft-versus-host disease; ICD = International Classification of Diseases; MERS = Middle East respiratory syndrome; MODS = multiple organ dysfunction syndrome; PRES = posterior reversible encephalopathy syndrome; SARS = severe acute respiratory syndrome; SIRS = severe inflammatory response syndrome; SSSS = staphylococcal scald skin syndrome; TLS = tumour lysis syndrome; TMA = thrombotic microangiopathy; TSS = toxic shock syndrome; VOD = veno-occlusive disease
Some of these outbreaks were not ARDS, making them unnecessary and potentially a form of ARDS or atypical pneumonia causing MERS. These acronyms (SARI and MERS) are widely used but essentially represent medical intervention to achieve homeostasis. The acronym syndromes encountered in CCM settings are descriptive terminologies that should remain simple in definition. However, such acronyms in the recent COVID-19 pandemic due to the virus SARS-CoV-2. Paediatric multisystem inflammatory syndrome (PMIS), multisystem inflammatory syndrome in children (MIS-C), and paediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2 (PIM-TS) are all recently coined acronyms for a systemic disease involving persistent fever, inflammation, and organ dysfunction following exposure to SARS-CoV-2. This syndrome has been considered to resemble Kawasaki disease and SIRS and associated with the hyperinflammation in cytokine release syndrome and cytokine storm syndrome.

**Conclusion**

The acronym syndromes encountered in CCM settings are descriptive terminologies that should remain simple in definition. However, such
syndromes may not necessarily aid diagnosis and prognostication. Users of these acronyms must understand the limitations and confusions behind these terminologies. Terms such as SARI, ALI, TSS, and SSSS are used infrequently and should be eliminated to avoid confusion. The underlying aetiologies of any syndrome must be explored and treated. For disease coding and prognostication analyses, all relevant aetiological factors must be considered.

**Author contributions**

All authors contributed to the concept of the study, acquisition and analysis of the data, drafting of the manuscript, and critical revision of the manuscript for important intellectual content. All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

**Conflicts of interest**

As an editor of the journal, KL Hon was not involved in the peer review process. Other authors declare that they have no other conflict of interest.

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**References**