Formulation and Reporting of Guidelines: Providing More Information Will Make Them Better

The recently published consensus guidelines for immunization of children with cancer in India [1] address an important 'felt-need' of the practicing pediatricians. It has been stated in the article that these guidelines were in the making since 2014 and were finalized in 2018. Based on the list of references cited, it appears that although the experts have considered various guidelines published till 2018, they have only reviewed original data generated till 2014. Since the guidelines have been published in December 2019, it would have been better if original studies, especially the Indian ones, published till 2019 were reviewed and considered while making the recommendations. It was also desirable to provide information regarding the quality of evidence supporting every recommendation and the strength of each recommendation. This would have helped healthcare providers in making appropriate informed decisions with greater confidence. It may be noted that the Infectious Disease Society of America guidelines that the authors have referred to, provide such information for each recommendation [2].

In the interest of enhancing confidence in such guidelines, it is necessary that the experts formulating these provide greater details of the methodology used for arriving at the recommendations (including the search strategy and process, evidence selection criteria, process of evaluation of the quality of evidence, procedure for formulating recommendations, use of external review and quality assurance process). A gist of these procedural details can be published in the published guidelines, and comprehensive methodological details can be put up at the organization's website. This will enhance the transparency in the system of making recommendations and will also allow a critical appraisal of the judgments made while formulating the guidelines [3].

With a view to ensure comprehensive reporting, the editors of *Indian Pediatrics* encourage authors to adhere to relevant guidelines (*eg*. CONSORT guidelines and STROBE guidelines, etc) while writing research articles [4]. They can extend this principle to reporting recommendations and practice guidelines by encouraging the expert groups to adhere to AGREE Checklist [5] or the RIGHT statement [6], while formulating and reporting recommendations. This will promote transparency, allow critical appraisal and assure the readers about the evidence-base of the recommendations made.

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AUTHORS' REPLY

We thank the reader for his interest in the guideline on immunization of children treated with chemotherapy [1]. We agree that five years (2014-2019) was a long time for this guideline to develop from inception of the idea to publication; however, despite the lag time, the published version was updated immediately pre-publication in line with the Indian Academy of Pediatrics (IAP) Advisory Committee on Vaccines and Immunization Practices (ACVIP) 2018-2019 immunization recommendations [2] (which in itself is updated based on current evidence) based on pre-approval review by the ACVIP itself.

We also acknowledge that it is essential to detail the methodology of guideline development and grade the strength of recommendations based on the level of evidence. A companion article with more detailed review of literature and gradation of available evidence was previously submitted to this journal along with the guidelines, and is currently under review for publication in a different journal. When considered together, the companion article should be able to complement the published guidelines, so as to fulfill many of the items mentioned in the AGREE 2016 checklist [3].

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Mauriac Syndrome in a Young Child with Diabetes

Mauriac syndrome is a rare complication of type 1 diabetes mellitus, usually reported in adolescents with poor glycemic control. It is characterized by hepatomegaly due to glycogen deposition, growth failure and delayed puberty [1]. Often these children have cushingoid facies, elevated liver enzymes and dyslipidemia in the form of increased cholesterol and triglyceride level in blood [2].

A two-year-old girl, diagnosed with type 1 diabetes six months back, presented with severe diabetic ketoacidosis. She had been advised split-mix regime but had poor compliance with the treatment. On examination, she had tachypnea, growth failure (weight and height <-2 SD for age), cushingoid facies, distended abdomen, and hepatomegaly (span 10 cm).

Investigations showed elevated blood glucose of 506 mg/dL, severe ketoacidosis (blood pH 6.98) ketonuria, HbA1c 11.5g/dL, high serum triglycerides (207mg/dL) and serum total cholesterol levels (192 mg/dL), elevated hepatic transaminase (AST 183 U/L, ALT 196 U/L), and normal antinuclear antibodies, thyroid function tests and anti-tissue transglutaminase levels. Ultrasound of abdomen showed normal echotexture of liver and normal intrahepatic biliary redicles. Ketoacidosis was managed as per standard protocol, and she was discharged on subcutaneous isophane and regular insulin. There was normalization of hepatomegaly and elevated liver enzymes after two months and liver biopsy was deferred. Mauriac syndrome was considered as the most probable explanation for liver dysfunction in this child.

There are anecdotal case reports of mutation in *PHKG2*, which is the catalytic subunit of the enzyme glycogen phosphorylase kinase. Hepatomegaly is a cardinal feature of Mauriac syndrome present in the majority, which occurs due to hepatic glycogen deposition due to the facilitated glucose diffusion across the hepatocytes [3]. The possible mechanisms for growth failure in Mauriac syndrome are inadequate tissue glucose availability, reduced circulating IGF-I level, and a relative growth hormone-resistant state [4]. The cushingoid features probably occur due to secondary hyperadreno-corticism. Short acting insulin regimens and brittle glycemic control are predisposing factors in these children for this rare complication. Most children with Mauriac syndrome are reported during adolescence. To the best of our knowledge, the youngest case of this syndrome reported earlier was aged three years [5]. Clinicians should suspect it in any child with uncontrolled diabetes and hepatic dysfunction. Identification of this syndrome indicates poor disease control, which can guide further management.

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