Author’s response to reviews

Title: Complete Agenesis of the Dorsal Pancreas Presenting with Diabetic Ketoacidosis – A Case Report and Literature Review

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Thank you very much for your suggestions. Our responses and revisions are as follows.

1. Differently to what written, this is not the first study describing beta-cell function in patients with agenesis of the dorsal pancreas (see Caetano LA et al., Clin Genet. 2018 Feb;93(2):382-386). As well, there are reports of patients with a final diagnosis of ADP, who were initially treated as DKA (Devarbhavi et al., Annual Research &amp; Review in Biology, 4(16): 2579-2586, 2014). Nevertheless, since DKA is a rare presentation of ADP, this case report might still deserve to be published in BMC Endocrine disorder. Please refer to the above articles (which should be included in table 2) and modify the statements “there is no study on the involvement of diabetic ketoacidosis” and similar.

Response: Thank you for your careful review. We reviewed the articles suggested by the reviewer, and corrected our statements accordingly. The abstract and Table-2 were also updated.

Revision:
(1). However, there are only two studies reporting a correlation between ADP and DKA in English literature. (in section Abstract line31-32 page2 )
(2). Although β-cell dysfunction is often indicative of hyperglycemia, there are only two studies reporting a correlation between ADP and DKA [11, 12]. Four cases of ADP, including the present one, had reported C-peptide test results, three of which showed low levels of fasting and postprandial C-peptide associated with β-cell dysfunction [13, 14], and one case showed detectable C-peptide level of 0.47 nmol/L [11]. Therefore, low insulin levels underlie most of the glucose metabolism disorders, as islets and β-cells are located in the tail of the pancreas [15, 16]. Previous studies have reported variations in the severity of high-fasting blood glucose disorders and insulin-dependent diabetes [10, 17], indicating that there are many degrees of β-cell dysfunction in patients with ADP. (section Discussion and Conclusion line106-113 page5)
2. The genetic study is essential for a full description of the clinical case. Please add this as a limitation of this case report.
Response: Thank you for your suggestion. We have addressed this issue as a limitation in this report.
Revision: However, one limitation should be noted that we didn’t have genetic analysis in the presented case as the patient refused DNA sequencing test. (section Discussion and conclusion line 95-96 page4)

3. An evaluation of patients’ relatives should be performed.
Response: The family history was noncontributory. We described the medical history of the patient’s mother, father, and younger sister in detail.
Revision: His mother died of gynecological cancer at age 50. His father had no history of hyperglycemia or chronic abdominal pain, and the abdominal CT scan showed a normal pancreas. His only younger sister had no special medical history as well. (section Case Presentation line 58-60 Page3)