Author’s response to reviews

Title: Case Report of One month and 15 days old baby with Type V Hyperlipoproteinemia

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Editor-in-Chief

Dr. Aldo Ferreira-Hermosillo,

Subject: BEND-D-19-00409 R1

Title: Case Report of One month and 15 days old baby with Familial hyperchylomicronemia syndrome (FCS)

Dear Sir,

Thank you much for reviewing our case report. When we wrote and submit this case it was diagnosed type 1HLP, but later on we diagnosed as type 5HLP upon further workup.

All comments and suggestion have edited. All changes made are mentioned below

EDITOR COMMENTS:

The current clinical case requires additional information for supporting their FCS diagnosis.

1. Comment: Please provide more information regarding lab test: was serum diluted for biochemical analyses?

1. Response: yes Serum was diluted to report exact levels of TGs on physicians request as the analytical measuring range for TGs on Cobas c-311 analyzer is 8 mg/dl to 885 mg/dl. The manual serial dilution was done 1:50 and exact 31400 mg/dl level were reported. (Added in case presentation section, 3rd paragraph, line no 120 -123)
2. Comment Did the authors perform any other biochemical marker that supports their FCS diagnosis? If not, please discuss it in the proper section

2. Response: Yes, lipoprotein electrophoresis was done on first follow-up visit it showed elevated Chylomicrons 4.7% (0-2%) and pre-beta lipoprotein or VLDL were 51.5 % (5-22 %) hence diagnosis of type 5 HLP was made. Table no 3 added (Added in case presentation section, 4th paragraph,lineno158-160,tablelineno177-183)

3).Comment How HDL and LDL were determined? The values of “05” and “02” reported in Table 1 at 3 months since admission to the hospital are correct?

3. Response: Yes these levels are correct patient is taking Cholesystramine 100mg/kg on t.i.d basis. HDL was performed by Homogeneous enzymatic colorimetric test on Cobas c-311 analyzer it Analytical Measurement Range (AMR) of 3.09-150 mg/dL, and LDL AMR 2-548 mg/dL was done by Homogeneous enzymatic colorimetric assay on Cobas c-311 analyzer(Added in case presentation section, paragraph 3rd, line no 123-125)

4).Comment: Please provide more information on dietary and medical treatment prescribed.

4. Response: Cholesystramine (Questran sachet) powder was started at dose of 100 mg/kg on t.i.d basis with NAN 1 formula Milk at the age of 1 month and 15 days and on follow up visit detailed weaning food advices given to mother (use olive oil during cooking of weaning food, give protein avoid fatty meal was advised. (Added in case presentation section, 4th paragraph, line 160-163)

REVIEWER 1 COMMENTS:

1. Comment: It’s seamed that the serum TG levels was measured with diluted sample. It was rare that serum triglyceride level was 31,400 mg/dl. Therefore, authors should describe more details data of another dyslipidemia marker (for example LPL activity, RLP-Cholesterol, ApoII-C and lipoprotein fractions etc.) for diagnosis of FCS without genetic testing.

In addition, please provide laboratory data (peripheral blood, blood biochemistry and urinary test)withanotherTable.
1. Response: yes serum was diluted to report the exact levels of TGs as analytical measuring range (AMR) for TGs on Cobas c-311 analyzer is 8 mg/dl to 885 mg/dl, so serial manual dilutions were done 1:50 (added in case presentation section, 3rd Paragraph, line 120-123).

Genetic testing like Apolipoproteins B (Apo B), ApoA-V, lipoprotein(a) Lp(a), ApoC-11 and Glycosylphosphatidylinositol-anchored high-density lipoprotein binding protein 1(GPIHBP1) could not done because of unavailability of these tests in our laboratory and even not available in our country and patients parents was not agree because of financial limitation. (Added in case presentation section, 4th paragraph, line 152-156)

A table made upon reviewer advice of available routine laboratory tests (added in case presentation, line no 130-147 as table no 1)

2. Comment In many case, primary FCS were caused by disorder of metabolisms of Chylomicrons. Therefore, Chylomicrons metabolisms markers of subject with FCS were deficiency or decreased even improved and/or decreased triglyceride levels.

Another did not perform genetic test, therefore they should describe that point with limitation, and provide current Chylomicrons metabolisms markers for diagnosis of primary FCS.

2. Response Genetic testing like Apolipoproteins B (Apo B), ApoA-V, lipoprotein(a) Lp(a), ApoC-11 and Glycosylphosphatidylinositol-anchored high-density lipoprotein binding protein 1(GPIHBP1) could not done because of unavailability of these tests in our laboratory and even not available in our country. (Added in discussion and conclusion section as limitation, 5th paragraph, line 219-221)

3. Comment: The patient was dramatically improved triglyceride level at 2 months. Did triglyceride level improve after Cholesystramine therapy? It was not clear the relation improved triglyceride and therapy.

Authors should describe more details for education of Dietary Modification and describe more details the Cholesystramine therapy (from when Cholesystramine therapy was started etc.).

3. Response. The patient was started Cholesystramine therapy at the age of 1 month and 15 days at that time he was on milk only so, he was suggested NAN-1 formula milk his lipid profile was repeated at age of 3 months at this time mother was advice for weaning diet to use olive oil in cooking weaning and advice give him protein diet like fish chicken, vegetables avoid fatty meals. (Added in case presentation section, 4th paragraph, line 160-163).
4. Comment: On DISCUSSION AND CONCLUSION session (line 208-217), the authors describe similar case reported. However, these reports were very severe hypertriglyceridemia which they did not perform genetic test. I think that most important point of this case reports were very severe hypertriglyceridemia caused in infant and improved with Dietary Modification and Cholesystraminetherapy.

Please discuss this point on DISCUSSION AND CONCLUSION session.

4. Response: we did not perform genetic testing because of unavailability of these tests in our country and patients’ parents were not agreeing to do from any other country due to financial limitation and he is doing with Cholestyramine therapy. he was advice use olive oil in weaning diet and advice to use protein diet avoid heavy fatty meal, his endocrinologist have planned to start statins in case if lipid profile will worse in future. (as per reviewer suggestion, added in discussion and conclusion section, paragraph, line no 219-221)

REVIEWER 2 COMMENTS:

1. Comment: Several words without separation among punctuation signs or words should be having corrections.

1. Response: correction done

2. Comment: 109-110 lines word space corrections

2. Response: correction done