ApoPharma Inc.

Data Safety Monitoring Board Communication

Date: 09 September 2014

To: TIRCON2012V1 study investigators (sites 001, 002, 003, 004)

From: Dr. Anna Rozova, Director, Medical Safety, ApoPharma Inc.

Re: Clinical study protocol, TIRCON2012V1: A randomized, double-blind, placebo controlled trial of deferiprone in patients with pantothenate-kinase associated neurodegeneration (PKAN)

DSMB safety review meeting of 12 August 2014

The third interim review meeting of the independent Data Safety Monitoring Board (DSMB) for the clinical study, TIRCON2012V1, was held on the 12th of August 2014.

The meeting was scheduled and held according to the planned periodic review of safety data. The review encompassed the safety data collected from the beginning of the study until the cut-off period of 09 July 2014 and any late-breaking safety information received between the cut-off point and the date of the meeting.

The DSMB reviewed the above-referenced data. The DSMB raised a concern about three (3) patients who show a decreasing trend in mean corpuscular volume (MCV), serum ferritin (SF) and haemoglobin (Hb). The DSMB conveyed the following request and recommendation:

- Obtain additional information on the three patients with the decreasing trend in MCV, Hb and SF to determine alternate cause(s) of anemia (affected sites will be contacted separately).
- If there is no additional explanation for the anemia in the specified patients, consider establishing guidelines such as: If Hb drops >3 g/dL from baseline and/or if absolute Hb value drops to less than 10 g/dL, a reduction in dose should be considered. Haemoglobin levels should be re-measured within one month.

The DSMB did not identify any issues requiring premature unblinding, suspension or termination of the study, and recommended the continuation of the study.