**Lipid clinic and lipoprotein disorders service (COVID-19\*)**

**Document 4: Volanesorsen subcutaneous injection
300mg weekly or fortnightly**

Rationale for administration of Volanesorsen:

* Familial Chylomicronaemia Syndrome (FCS)

Criteria for referral for Volanesorsen:

* Confirmation of FCS by genetic testing
* Recurrent history of acute pancreatitis
* Triglyceride levels above 10 mmol/L
* Platelets at baseline above 140 x10ꝰ/L

Procedure for Volanesorsen treatment:

* Above criteria met and blueteq form completed. Currently on Early Access to Medicines Scheme (EAMS).
* Informed consent taken from the patient to enter EAMS. Education and training for subcutaneous injections given. Patient alert card given.
* Dietetic input – advise very low fat eating plan 10-20g fat per day, requires ongoing support.
* Blood tests weekly or fortnightly dependent on platelet results (see below). These can be done by the GP also.
* Attend hospital monthly to pick up injections (only 2 injections supplied at any one time and only delivery to the hospital).

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| **PLT Level (count** x 109/L**)** | **Dose schedule** | **PLT Monitoring** |
| **Body Weight < 70 kg** | **Body Weight ≥ 70 kg** |
| **Normal (>140)** | Every 2 weeks | Every 2 weeks | Weekly+ | Every 2 weeks |
| **100-140** | Every 2 weeks | Every 2 weeks | Weekly+ | Weekly until stable |
| **75-100** | Pause, resume every 2 weeks when >100 x 109/L | Every 2 weeks+ | Weekly |
| **50-75** | Pause, resume every 2 weeks when >100 x 109/L | Twice per week until stable |
| **<50** | Discontinue volanesorsen\*\* | Every other day until stable\* |

+For patients up-titrated to a dose of 300 mg once weekly

\*\*Daily if PLT <25 x 109/L/mm3; if platelet count <25 x 109/L steroid therapy should be considered

\*\*For any patient dose paused or discontinued due to severe thrombocytopenia, the benefits and risks of returning to treatment should be carefully considered. For discontinued patients, a haematologist should be consulted prior to resuming treatment

\*Possible changes/adaptations because of COVID-19 epidemic (reasonable but not ideal):

* Should continue monitoring as usual because of risk of significant complications
* Try to coordinate blood tests in our outpatients
* Change face-to-face clinic appointments to telephone appointments unless if absolutely necessary
* Continue prescriptions and drug home delivery as usual
* Before each visit (if necessary) to make sure patients (and households) have no symptoms to suggest COVID-19.
* Advice to patients: DO NOT change or stop your regular medications without discussion with your consultant or specialist nurse.

Possible main side effects from Volanesorsen:

* Low platelets which may result in thrombocytopenia.
* Injection site reactions.
* Fatigue.
* Myalgia.
* Headaches.

Benefits of Volanesorsen:

* Reduces fasting triglycerides, total cholesterol, non-HDL cholesterol, apoC-III, apoB-48, and chylomicron triglyceride levels and increased LDL-C, HDL-C, and apoB100 (APPROACH study).
* Reduces episodes of acute pancreatitis and hospitalisations related to pancreatitis (pooled data from different studies).

Rationale for patients attending hospital for Volanesorsen follow up:

* Treatment requires close specialist supervision and monitoring.
* Secondary care prescription only in few selected centres in UK with experience in managing FCS and treatment with Volanesorsen.
* Regular dietetic support.