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Title:

Guidance for FVIII and FIX Inhibitor Testing

Abstract:

The major complication of haemophilia therapy today is the development of anti-drug (anti-factor) antibodies termed inhibitors. In severe haemophilia A, Factor VIII (FVIII) inhibitors form in approximately 30% of patients, usually during the first 20–30 days of CFC exposure, but in severe haemophilia B, the cumulative incidence of Factor IX (FIX) inhibitor development is lower than in severe haemophilia A and is as high as 4–5% after a median of only 9–11 exposure days.

Detection and monitoring of FVIII and FIX inhibitors are essential to patient management.

The assay methodology for FVIII and FIX inhibitors is rather complex and a number of variables may influence the test result. The complexity of the assay system may result in undesirably high within- and between-laboratory variation in test results. Considerable between-laboratory variation for FVIII inhibitor testing (30 – 60%) has been shown by several providers of external quality assessment programs. These observations demonstrate the lack of standardization for FVIII and FIX inhibitor testing.

The International Council for Standardization in Haematology (ICSH) has decided to draft a laboratory guideline for testing of FVIII and FIX Inhibitors.

The guideline includes the following aspects: screening for inhibitors, assay principle, sample requirements, testing requirements and interpretation, quality assurance, interferences and recent developments.

During the presentation the guideline will be presented and the major recommendations for standardized testing of FVIII and FIX inhibitor testing will be highlighted.