

Three problems of hemophilia B : a study of abnormal factor IX molecules with an inhibitor neutralization assay Briët, E.

Citation

Briët, E. (1977, June 16). *Three problems of hemophilia B : a study of abnormal factor IX molecules with an inhibitor neutralization assay*. Drukkerij "Luctor et emergo", Leiden. Retrieved from https://hdl.handle.net/1887/61512

Version:	Not Applicable (or Unknown)
License:	<u>Licence agreement concerning inclusion of doctoral thesis in the</u> <u>Institutional Repository of the University of Leiden</u>
Downloaded from:	https://hdl.handle.net/1887/61512

Note: To cite this publication please use the final published version (if applicable).

Cover Page



Universiteit Leiden



The handle <u>http://hdl.handle.net/1887/61512</u> holds various files of this Leiden University dissertation

Author: Briët, Ernest Title: Three problems of hemophilia B : a study of abnormal factor IX molecules with an inhibitor neutralization assay Date: 1977-06-16

GLOSSARY OF TERMS AND ABBREVIATIONS

٠

CRM-negative	_	plasma in which the immunologically esti- mated level of a protein and the level of its biological activity are equal.
CRM-positive	_	plasma in which a significant excess is found of the immunologically estimated level of a protein over the level of its biological activ- ity.
factor IX activity		clotting factor IX measured by its biological activity in the blood clotting mechanism.
factor IX antigen	—	factor IX protein measured by an im- munological assay, is therefore a synonym for factor IX-CRM i.e., material that pro- duces a cross-reaction with antibodies against factor IX.
factor IX inhibitor	_	antibody against factor IX present in the plasma of approximately 5% of patients with severe hemophilia B after treatment with factor IX transfusions.
hemophilia B—	_	variant of hemophilia B in which the plasma is CRM-negative.
hemophilia B+		variant of hemophilia B in which the plasma is CRM-positive.
hemophilia B Leyden	_	variant of hemophilia B in which the bleeding symptoms disappear after puberty along with a gradual increase of both factor IX activity and antigen level.
hemophilia B _M		variant of hemophilia B^+ in which a pro- longed ox-brain thromboplastin-time is found.

INA	—	inhibitor	neutralization	assay.	

- obligatory hemophilia carrier a women who is definitely a carrier of hemophilia on the basis of her family history i.e.: a daughter of a hemophiliac or a woman with hemophilic relatives who has at least one hemophilic son, or a woman who has two or more hemophilic sons.
- PIVKA Protein Induced by Vitamen K Absence (or in the presence of Vitamin K Antagonist); biologically inactive precursor of the vitamin K dependent clotting factors (II, VII, IX, X) that can be demonstrated by immunological assays.

ACKNOWLEDGEMENTS

I am indebted to Mr N. H. van Tilburg for his technical assistance, to Miss M. Heemskerk for preparing the manuscript, to Mr J. J. Magdelijns and Mr J. van Leeuwen for drawing the figures, to Mr. J. G. P. Tijssen and Mr J. Hermans for their statistical advice, to Dr K. H. Ørstavik (Oslo, Norway) for the electro-immuno assays of factor IX, to Dr H. R. Roberts (Chapel Hill, N.C., USA) who kindly provided the human factor IX inhibitor, to Dr R. M. Bertina, Dr J. G. Eernisse and Dr J. M. Sepers for the stimulating discussions, and to all the patients and their families who have been involved in this study.