INTENDED USE
The Factor IX Deficient Substrate Plasma is intended for the quantitative determination of Factor IX (Christmas factor) in patients suspected of having a congenital or acquired deficiency of this coagulation protein.

SUMMARY
Numerous coagulation factors have been identified in the blood and are required for normal blood clotting. A deficiency of one or more of these factors may result in a notable hemorrhagic condition, the severity of which is governed by the degree of the deficiency. Deficiencies of the blood clotting factors may be congenital or acquired. The congenital deficiencies are, in general, single deficiency states while the acquired deficiencies may be multiple and commonly associated with liver disease, vitamin K deficiency or the ingestion of coumarin type anticoagulant drugs, and defibrination secondary to intravascular clotting.

Factor IX, known as plasma thromboplastin component (FPC), is decreased in a congenital disease known as Hemophilia B or Christmas Disease. 1 Hemophilia B is clinically indistinguishable from hemophilia A, and it has a sex-linked recessive mode of inheritance. There are no known cases of hemophilia A to every case of hemophilia B. An acquired Factor IX deficiency may occur in conjunction with a vitamin K deficiency and/or hepatoellular dysfunction.2

In an effort to devise a quantitative assay for Factor IX, several methods based on the thromboplastin test were used and were found to be time consuming and complicated. Langstbel, Wagner and Blankenhorn (1963) developed a one-stage "partial thromboplastin time" which was simple to perform but not reproducible. Helena's procedure determines Factor IX activity by using a modification of the activated partial thromboplastin time (APTT) test and a Factor IX deficient substrate plasma.

PRINCIPLES
Quantitative measurement of individual coagulation factors by the one stage method depends upon having a substrate plasma lacking the factor being measured. A severely depressed substrate plasma (less than 1% activity) has a prolonged activated partial thromboplastin time (APTT). A dilution of the test plasma is mixed with an equal volume of factor deficient plasma, and the clotting time is determined before and after correction provided by the test plasma with the correction obtained from an accepted known reference plasma, the percent activity of the coagulation factor may be determined.3

REAGENT
Factor IX Deficient Substrate Plasma (Cat. No. 5194) Ingredients: The Factor IX deficient plasma contains less than 1% Factor IX activity.

Precautions: For Diagnostic Use Only. Avoid Ingestion. The Factor IX Deficient Substrate Plasma has been found to be positive when tested for Hepatitis B Antigen (HBeAg) and HIV antibody; however, the deficient plasma should be handled with the same precautions as those observed when working with patient's blood.

Storage and Stability
The optimized product is stable until the expiration date printed on the vial and box labels when stored at 2 to 6 °C. The reconstituted product is stable for 8 hours at 2-8 °C. After reconstitution, the product should be kept on ice for the duration of testing.

Shrinkage indication that test has been modified, added or deleted.

INDEX
1. Assay patient samples as soon after collection as possible.
2. Sample dilutions must be assayed within 30 minutes after preparation and maintained on ice until tested.
3. Sample dilutions exceeding 1:40 are not recommended.
4. Do not re-use the recommended dilutions on plasma samples to avoid erroneous results due to possible dilution errors.
5. When performing Factor assay, more than one vial of reagent may be needed. To eliminate vial-to-vial variation multiple vials should be reconstituted, allowed to dissolve, and then pooled.

Shade enclosed indicates that text has been modified, added or deleted.
INTENDED USE
The Factor IX Deficient Substrate Plasma is intended for the quantitative determination of Factor IX (Christmas factor) in patient suspected of having a congenital or acquired deficiency of this coagulation protein.

SUMMARY
Coagulation factor deficiencies have been identified in the blood and are responsible for the bleeding tendency associated with the deficiency. Deficiencies of the blood clotting factors may be congenital or acquired. The congenital deficiencies are, in general, single factor deficiencies whereas the acquired deficiencies may be multiple in nature and commonly associated with liver disease, vitamin K deficiency or the ingestion of coumarin type anticoagulant drugs, and defibrinogenation secondary to intravascular clotting.

Factor IX, known as plasma thromboplastin component (PTC), is decreased in a congenital disease known as Hemophilia B or Christmas Disease. 1 Hemophilia B is clinically indistinguishable from hemophilia A, and it has a sex-linked recessive mode of inheritance. There are several cases of hemophilia A to every case of hemophilia B.

A deficiency of Factor IX may occur in conjunction with a vitamin K deficiency and/or hepatocellular dysfunction. 1 In an effort to devise a quantitative assay for Factor IX, several methods based on the thromboplastin test were used and were found to be time consuming and complicated. Langleit, Wagner and Brinkhaus (1953) developed a one-stage “partial thromboplastin time” which was simple to perform but not reproducible. Helena’s procedure determines Factor IX activity by using a modification of the activated partial thromboplastin time (aPTT) test and a Factor IX deficient substrate plasma.

QUALITY CONTROL
A. Specimen and Reagent Preparation
Factor IX has a decreased activity in a congenital condition known as hemophilia B or Christmas Disease, which is sex-linked recessive. One type of this disease is caused by an abnormal condition known as S.B.C. 2 . The Factor IX Deficient Substrate Plasma is limited to Factor IX activity determinations based on a modified APTT test system. Dilutions of the test specimen exceeding 1:40 are not recommended. For the amount of reagent factor X required, Helena’s Coagulation S.A.R.P. (Cat. No. 5185) is recommended for use as the standard.

B. Standard Curve Preparation
Preparation for the procedure, all test tubes, syringes, and materials must be plastic or siliconized glass 2 .

C. Procedure
1. Assay patient samples as soon after collection as possible.
2. Sample dilutions must be assayed within 30 minutes after preparation and maintained on ice until tested.
3. Sample dilutions exceeding 1:40 are not recommended.
4. The results of the recommended dilutions on plasma samples should be reconstituted, allowed to dissolve, and then pooled. 1

Signs of Deterioration
The lyophilized product is stable until 10 x 5.0 mL - 250 tests 5383 10 x 10 mL - 500 tests 5384 10 x 10 mL - APTT Reagent 5385 10 x 10 mL - Calcium Chloride 5386

Materials Required but not provided:

- 12 x 75 mm plastic test tubes
- Stopwatch
- Deionized water


tube. Swirl gently and allow to stand for 10 minutes to ensure complete dissolution.

Factor IX Deficient Substrate Plasma (Cat. No. 5194) Ingredients: This is a human plasma which contains less than 1% Factor IX activity.

Precautions: Factor IX Diagnostic Use Only. Avoid ingestion. The Factor IX Deficient Substrate Plasma has been found negative when tested for Hepatitis B (Anti-HBsAg) and HIV antibody; however, the deficient plasma should be handled with the same precautions as those observed when handling whole blood.

Procedure for Reconstitution
Factor IX Deficient Substrate Plasma with 1.0 mL deionized water. Swirl gently and allow to stand 15 minutes at room temperature to ensure complete dissolution.

Storage and Stability
Prepared Factor IX deficient plasma is stable until the expiration date printed on the vial and box labels when stored at 2 to 6°C. The reconstituted product is stable for 8 hours at 2-6°C. After 8 hours, the sample should be discarded. The product should be kept on ice for the duration of testing.

Signs of Deterioration
The lyophilized product may appear as a dry, straw colored plug or pieces.