



GUIDELINES FOR THE TREATMENT OF CONGENITAL DISORDERS OF FIBRINOGEN

**Australian Haemophilia Centre Directors'
Organisation**

November 2010

Disclaimer

This document is a general guide to appropriate practice, to be followed subject to the clinician's judgment and the patient's preference in each individual case. The guidelines are designed to provide information to assist decision-making and are based on consensus opinion at the time of compilation (November 2010).

These guidelines will be reviewed in 2013.

Users can contact the Australian Haemophilia Centre Directors' Organisation by writing to:

1622 High Street,

Glen Iris

Victoria 3146

1. Introduction

Congenital disorders of fibrinogen are rare and may be classified as:

- Afibrinogenaemia
- Hypofibrinogenaemia
- Dysfibrinogenaemia

Clinical features include mucocutaenous bleeding and deep tissue bleeding including intracranial bleeding. Umbilical cord bleeding is a common presentation of congenital afibrinogenaemia. Paradoxically, arterial and venous thromboses have been reported in patients with disorders of fibrinogen. Genetic diagnosis is available (contact Prof. Stephen Brennan, Canterbury Health Laboratories, New Zealand, telephone +64 3 364 0549; email steve.brennan@otago.ac.nz)

The half life of fibrinogen is approximately 4 days and the normal plasma concentration is approximately 1.5 to 3.5g/l.

2. Treatment Products Available in Australia

2.1. Riastap (formerly Haemocomplettan P)

Riastap is purified concentrated fibrinogen derived from human plasma. Riastap is available in 2 vials sizes (1 gram vial contains 900 – 1300mg of human fibrinogen, 2 gram vial contains 1800 to 2600mg of human fibrinogen). Production of Riastap includes viral inactivation processes (pasteurisation (at +60°C for 20 hours) and multiple precipitation and absorption steps). Riastap is available via the Special Access Scheme.

2.2. Cryoprecipitate

Cryoprecipitate is issued by the Australian Red Cross Blood Service. This is a plasma derived blood product that does not undergo any viral inactivation process. One bag of cryoprecipitate contains at least 140mg of fibrinogen in a bag of 30 – 40ml.

3. Treatment Regimes

Surgical procedures should be planned and performed in conjunction with a haemophilia treatment centre. In principle the use of a specific concentrate which is virally inactivated is preferred. Cryoprecipitate is readily available and should be used in an urgent situation if Riastap is not available.

Recommended dosage for the treatment of bleeding episodes is according to the formula;

Dose (g) = desired increment (g/l) X plasma volume X weight (kilograms).
(plasma volume is 0.07 X (1-haematocrit))

The half life of fibrinogen is long (3 – 4 days). Depending on the clinical situation further doses may be required, the frequency of which should be based on monitoring of fibrinogen levels. In general the fibrinogen level to be achieved is 1g L which should then be maintained at this level until haemostasis is achieved and wound healing complete. Fibrinogen levels should be monitored regularly depending on the clinical indication or response.

Antifibrinolytic therapy may be used for minor bleeding episodes particularly for dental bleeding; caution should be exercised in any patients with a history of thrombosis or thrombotic disorders

Pregnancy is a particularly risky time for females with congenital fibrinogen disorders, including the potential for early fetal loss. During pregnancy, early treatment may be necessary which should be managed at a haemophilia treatment centre in conjunction with a specialist obstetric service.

4. Thrombotic Risk

There are case reports of thrombosis in patients with congenital fibrinogen disorders (including afibrinogenaemia). Caution should be exercised to prevent overtreatment.

Reference

de Moerloose P & Neerman-Arbez M, Congenital fibrinogen disorders, *Seminars in Thrombosis & Hemostasis*.2009 Jun, 35(4), 356-66.