

## Uso de rVIIa en pacientes con déficit de factor VII

Successful long term prophylaxis using activated recombinant factor VII (rVIIa) in a young girl with severe factor VII deficiency

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We report the case of a 30 month old girl, born full term from consanguineous parents after uncomplicated pregnancy. At the age of 3 days, she presented a haematemesis; the prothrombin time was dramatically prolonged and she was diagnosed with a severe FVII deficiency (FVII <3%). Two months later, she was admitted with a left partial comitial attack, secondary to a subdural haematoma with compression. She was placed immediately on mechanical ventilation, anti-epileptics and a substitution with rFVIIa at 35 mcg /kg was started every 3 h. Initially no surgical intervention was undertaken. At day 2, she presented partial left hemiplegia secondary to a cerebral infarct (sylvius territory). There was no evidence for thrombosis and surgery was undertaken. The rFVIIa was administered upon admission at 30mcg/kg every 3 h for 3 days, every 4 h for 2 days, every 6 h during mechanical ventilation time (45 days). During the thirty last days, syringes ready-for-use were filled by the pharmacist: 14mg of rFVIIa were used for 100 doses during 30 days. The total cost was 9216 euros versus 76800 euros if we had used the ready-to-use commercial presentation. After 3 months, a long term prophylaxis with rFVIIa at 30mcg/kg two times a week was initiated, which was performed at home since June 2005. Actually, she remains on twice a week prophylactic infusions without any evidence of new bleeds. In conclusion, we report a successful long term prophylactic treatment with rFVIIa in a severe FVII deficiency and the economic interest of rFVIIa small dosages.

Este poster presentado en el XXVIII Congreso Internacional de la World Federation of Hemophilia en Estambul en junio de 2008 ofrece otra evidencia de la eficacia del rFVIIa en el tratamiento profiláctico en el déficit severo de factor VII.

Una recién nacida debuta a los 3 días de vida con hematemesis y en ese momento se hace diagnóstico de déficit severo de factor VII. A los 2 meses de vida presenta convulsiones secundarias a hematoma subdural que requirió cirugía y que se manejó con rFVIIa con intervalos crecientes. Dada la severidad de la sintomatología y lo temprano de la aparición del sangrado, la paciente justificaba claramente el tratamiento profiláctico. El bajo peso de la paciente planteó un problema, especialmente económico, ya que usando las presentaciones comerciales habituales del rFVIIa se desperdiciaba parte del concentrado. La adaptación a dosis más pequeñas, preparadas por el farmacéutico, permitió un ahorro importante.

La profilaxis con rFVIIa dos veces por semana evitó nuevos sangrados y es una conducta que debe considerarse en deficiencias severas de factor VII con clínica florida de hemorragia y peligro de vida.