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A severe hemorrhagic diathesis has been discovered in a 10 years old girl whose brother has severe hemophilia with classical haemarthrosis. The factor VIII biological activity is less than 1% in both of them with an almost normal factor VIII related antigen. The bleeding time was normal. There was no consanguinity and paternity was not disproved by extensive blood grouping tests. Cytogenetic studies showed a deletion of a part of the long arm of one X chromosome. The formula of the karyotype is 48, X, del X (q. 21). The sex chromatin showed the presence of a phenotypic female. The X chromosome was smaller than normal. This is the first case to our knowledge of true female haemophilia A due to a deletion of one X chromosome.


Growth hormone (GH) has been implicated in the pathogenesis of diabetic angiopathy although the exact mechanism remains unknown. von Willebrand factor activity (vWF) significantly higher in diabetic than in normal plasma (p<0.001). In view of this, we have investigated the relationship of radioimmunoassayable GH and vWF activity, we measured in a biocentrone using washed normal human platelets and ristocetin. Samples were obtained after an overnight fast, during oral glucose tolerance tests, and during sleep. A significant positive correlation was seen between GH over a range of 0.46 to 12.3 mg/ml and vWF activity, we measured in a biocentrone using washed normal human platelets and ristocetin. Samples were obtained after an overnight fast, during oral glucose tolerance tests, and during sleep. A significant positive correlation was seen between GH over a range of 0.46 to 12.3 mg/ml and vWF activity. vWF activity was suppressed by oral glucose in normals and diabetics, with the amount of suppression related to the degree of glucose intolerance. Maximal suppression of vWF activity was coincident with maximal GH suppression. Samples from 4 normal subjects and from 4 normal subjects and from 4 normal subjects and from 4 normal subjects and from 4 normal subjects and from 4 normal subjects and from 4 normal subjects and from 4 normal subjects and from 4 normal subjects and from 4 normal subjects and from 4 normal subjects and from 4 normal subjects and from 4 normal subjects and from 40-minute intervals during sleep showed peaks of plasma GH and vWF activity with GH peaks preceding vWF peaks by an hour or less. Two patients with hypoglycemia and one patient with isolated GH deficiency were similarly studied. GH levels were low in these patients, but vWF levels were normal; both GH and vWF showed little sleep-related change. A single intramuscular injection of GH produced a marked increase of vWF activity in all 3 patients within 30 minutes and a later increase after 5-6 hours. We conclude that a regulator of glucose metabolism, possibly GH, is involved in regulation of vWF activity.