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WOLFRAM SYNDROME. PRESERVATION OF ADH SECRETION
THROUGH BARORECEPTORS WITH NO RESPONSE TO OSMORECEPTOR
STIMULATION.

Girl, aged 14 years, developed diabetes mellitus at the age of 2.5. At age 10.5 atonia of the urinary tract and bladder was diagnosed. She also had high frequency hearing loss and impaired vision (2/10 bilaterally) due to optic atrophy. No angiopathy of the retina. There was increased urinary volume with adequate diabetic control (7-9 L per 24 hours), and normal values of BUN, serum creatinine, creatinine clearance, and DMSA. Plasma osmolality (P.O.) was 307, 313, 310, with corresponding urine osmolality (U.O.) values of 142, 162, 158 mOsm/L H₂O respec-

tively, and plasma ADH (by RIA) 0-1 pg/ml. Plasma Renin activity and aldosterone values were within normal limits (1.1 to 2.2/ ng/ml/hr and 5.3 to 17.4 ng/dl respectively). In one deprivation test, P.O. was 310 mOsm/L H₂O, and U.O. 159 mOsm/L H₂O, while ADH was 0 pg/ml. However in a fainting spell which occurred during a water deprivation test and was associated with severe, momentary drop of blood pressure, plasma ADH rose to 9 pg/ml. The findings are suggestive of a dichotomy in the impairment of ADH secretion, analogous to the one reported in hypothalamic hypernatremia-hypodipsia syndrome; Namely normal ADH secretion through baroreceptors but impaired secretion through osmoreceptors. It is likely that in Wolfram syndrome transmission of the stimulus through osmoreceptors rather than deficient synthesis of ADH is at fault.