

at age 22 yr, which was attributed to pneumonia without associated renal failure. Dilated urinary tracts without diabetes insipidus were found by Blasi et al. (6) in more than 6% of their series of 168 patients collected from the literature. Patient longevity was not discussed by them or Fishman (7).

In a previously reported exhaustive survey of American endocrinologists, Swift et al. (8) were able to collect only 68 case reports they accepted as bona fide Wolfram's. At the time their survey ended in 1986, no living patients with Wolfram's syndrome older than 39 yr had been reported to them and only 2 were known to have lived past 40 yr. Lim and Thai (9) recently listed the varieties of reported urinary tract abnormalities in Wolfram's patients.

Because of its early onset, many of the reports and discussions of patients with Wolfram's syndrome have been printed in the pediatric and ophthalmologic literature. Recently, when an adult patient of ours in her early thirties with Wolfram's syndrome, which had been diagnosed in childhood, was hospitalized in our medical ICU, we found that our usually knowledgeable medical residents, as well as many of our attendings, were unfamiliar with this fascinating but unusual syndrome. Because of the importance of recognizing that genitourinary factors other than diabetes insipidus and diabetic nephropathy may contribute to the renal failure in these unfortunate patients, it becomes even more important to identify the syndrome. At the same time, smaller families make identification of recessive problems more difficult.

Wolfram patients are now likely to live long enough to become internists' rather than pediatricians' patients. The presence of renal tract abnormalities needs to be periodically assessed; renal failure should not be attributed to diabetes per se.

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HLA, HUMAN LEUKOCYTE ANTIGEN; IDDM, INSULIN-DEPENDENT DIABETES MELLITUS.

References

1. Wolfram DJ: Diabetes mellitus and simple optic atrophy among siblings: a report of four cases. *Mayo Clinic Proc* 110:715-18, 1938
2. Pilley SFJ, Thompson H: Familial syndrome of diabetes insipidus, diabetes mellitus, optic atrophy, and deafness (DIDMOAD) in childhood. *Br J Ophthalmol* 66:294-98, 1976
3. van Haefen TW, Razenberg PP: DIDMOAD syndrome and HLA-DR haplotype. *Horm Metab Res* 21:548-49, 1989
4. Marquardt JL, Loriaux DL: Diabetes mellitus and optic atrophy with associated findings of diabetes insipidus and neurosensory hearing loss in two siblings. *Arch Intern Med* 134:32-37, 1974
5. Khardori K, Stephens JW, Page OC, Dow RS: Diabetes mellitus and optic atrophy in two siblings: a report on a new association and a review of the literature. *Diabetes Care* 6:67-70, 1983
6. Blasi C, Pierelli F, Rispoli E, Saponara M, Vingolo E, Andreani D: Wolfram's syndrome: a clinical, diagnostic, and interpretative contribution. *Diabetes Care* 9:521-28, 1986
7. Fishman L, Ehrlich RM: Wolfram Syndrome: report of four new cases and a review of the literature. *Diabetes Care* 6:405-408, 1986
8. Swift RG, Sadler DB, Swift M: Psychiatric findings in Wolfram Syndrome. *Lancet* 336:667-69, 1990
9. Lim MCL, Thai AC: A Chinese family with Wolfram syndrome presenting with rapidly progressing diabetic retinopathy and renal failure. *Ann Acad Med Singapore* 19:548-55, 1990

Decision Support Systems for Diabetes Management

The term "Decision Support System" caught my attention in the recent article by Peters et al. (1). However, after reading the article, I was disappointed that little explicit consideration was given to the nature and setting of the decisions that were being supported. My understanding of current thinking in the literature of decision support systems is that it centers on issues such as the analysis and decomposition of the cognitive load imposed by the decision (2-3); the extent to which the decision is skill based, rule based, or knowledge based (4); and the importance of previous experience with such decisions.

I believe that research into the nature and effectiveness of insulin dosage recommendations to patients, whether given directly by health professionals or suggested by machines, would be usefully informed by this literature. For example, the explicit separation of decisions into skills (taking an injection), rules (given a blood glucose reading, what dose should you take), and knowledge (what do you do when you drop and smash your only bottle of insulin on a nonstop flight from San Francisco to Sydney) is a simple but informative way to think about decision support in diabetes management.

Further, I suggest that such research should focus on the identification of those aspects of the decision process or those situations in which human performance is somehow impaired and where computer assistance would be useful. This impairment may be the result of a lack of experience, an unusual meal situation, illness, or a hypoglycemic state. My current blood glucose meter signals me to check ketones when readings are >300 mg/dl. Perhaps future versions would both suggest an insulin dose (based on previous measurements) and shift into a more ver-