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.

ROBERT W. COX, MD

FROM THE MEDICAL CENTER OF DELAWARE, WILMINGTON, DELAWARE.

ADDRESS CORRESPONDENCE TO ROBERT COX, MD, OMEGA PROFESSIONAL CENTER, A94, NEWARK, DE 19713.

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 Haeften 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-

bose mode when a test result is below some threshold that suggests mental confusion.

E. BRUCE MACDOUGALL, PHD

FROM THE DEPARTMENT OF LANDSCAPE ARCHITECTURE AND REGIONAL PLANNING, UNIVERSITY OF MASSACHUSETTS AT AMHERST, AMHERST, MASSACHUSETTS.

ADDRESS CORRESPONDENCE TO E. BRUCE MACDOUGALL, PHD, DEPT. OF LANDSCAPE ARCHITECTURE AND REGIONAL PLANNING, UNIVERSITY OF MASSACHUSETTS AT AMHERST, HILLS NORTH, AMHERST, MA 01003.

References

1. Peters M, Rubsamen M, Jacob U, Look D, Scriba PC: Clinical evaluation of decision support system for insulin-dose adjustment in IDDM. *Diabetes Care* 14:875–80, 1991
2. Zachary WW: Decision support systems: designing to extend the cognitive limits. In *Handbook of Human-Computer Interaction*. Helander M, Ed. New York, Elsevier, 1988, p. 997–1030
3. Smith CL, Sage AP: A theory of situation assessment for decision support. *Inform Design Tech* 17:91–124, 1991
4. Rasmussen J: *Information Processing and Human-Machine Interaction*. New York, Elsevier, 1986

Lipid and Lipoprotein Levels in Young IDDM Patients

In a recent report of the DCCT Research Group (1), minor differences in lipid and lipoprotein levels were found comparing young (13–40 yr of age) IDDM volunteers with control values of the LRC program. Only in young females with relatively higher HbA_{1c} levels were elevated cholesterol, LDL-cholesterol, and TG values observed.

However, more profound differences in lipid levels between diabetic and nondiabetic populations have been noted in older studies. The authors (1) comment on this discrepancy—that a major change in dietary habits during the past 10–20 yr with a decrease in fat and increase in carbohydrate intake may play an important role.

In 1979, we investigated the lipid and lipoprotein levels in diabetic children (8–12 yr of age, $n = 30$) at a summer camp of 4-wk duration (2). Their caloric intake and dietary pattern was monitored by a dietian. The mean caloric intake was 2793 ± 814 cal/day, consisting of 40–45% fat, 20–25% protein, and 35–40% carbohydrate. The distribution of calories differed clearly from the current dietary advice and practice.

The mean HbA_{1c} was 9.8 ± 0.4% (normal range 4.6–6.6%); the mean insulin dosage was 0.8 ± 0.29 U/kg. Only on admission day did we observe increased total cholesterol ($P < 0.05$) and TG in females and elevated TG levels in males, compared with lipid levels of 64 healthy children. These differences were no longer apparent after 4 wk camping on the rather fat-rich diet described above; and total cholesterol, HDL cholesterol, LDL cholesterol, VLDL cholesterol, and TG were in the normal range for boys and girls.

This observation, although made among only a small group of patients, indicates that in otherwise healthy and normally active young diabetic patients with moderate metabolic control, lipid and lipoprotein levels are in the normal range. In accordance with the DCCT study, the evidence indicates that females may be more prone to alterations of the lipid levels in this age group. Changes in dietary habits may only partly explain the lower lipid levels reported by DCCT study (1).

E. SCHOBER, MD

FROM THE UNIVERSITY CHILDREN'S HOSPITAL, VIENNA, AUSTRIA.

ADDRESS CORRESPONDENCE TO E. SCHOBER, MD, UNIVERSITY CHILDREN'S HOSPITAL, WAHRINGER GURTEL 18–20, 1–1090 VIENNA, AUSTRIA.

DCCT, DIABETES CONTROL AND COMPLICATIONS TRIAL; IDDM, INSULIN-DEPENDENT DIABETES MELLITUS; LDL, LOW-DENSITY LIPOPROTEIN; TG, TRIGLYCERIDES; HDL, HIGH-DENSITY LIPOPROTEIN; VLDL, VERY-LOW-DENSITY LIPOPROTEIN.

References

1. The DCCT Research Group: Lipid and Lipoprotein Levels in Patients with IDDM: Diabetes Control and Complications Trial Experience. *Diabetes Care* 15:886–94, 1992
2. Pollak A, Widhalm K, Havelec L, Frisch H, Schober E: Glycosylated hemoglobin (HbA_{1c}) and plasma lipoproteins in juvenile onset diabetes mellitus. *Acta Paediatr Scand* 69:475–79, 1980

Response to Dr. Schober

We read with great interest the comments by E. Schober comparing the baseline lipid and lipoprotein measurements of the DCCT cohort (1) with lipid data from IDDM children attending a summer camp in 1979 (2). There are several notable differences between the DCCT study cohort and the population studied by Scrober et al. (2), including differences in nationality and period of study, and younger age (2–12 yr) in the Austrian study. Despite these differences, we consider the results with regard to lipid measurements to be mutually confirmatory. At baseline, the 19 Austrian children had lipid values that were similar to a nondiabetic population except for higher TG levels in the diabetic girls and boys. The diabetic girls had a slightly higher total cholesterol than the diabetic boys, although no significant difference was observed in total cholesterol between the diabetic and nondiabetic girls. Of note, a study by