

Biomedicine

Ingrid Wickelgren reports from Baltimore at the Johns Hopkins Centennial Science Writers Seminar

Drug shows promise in sickle cell anemia

Early clinical studies suggest that a drug normally used to treat leukemia may be effective in treating sickle cell anemia, a genetic disorder in which patients produce defective hemoglobin, the oxygen-carrying molecule within red blood cells. Scientists pinpointed the mutation responsible for sickle cell anemia more than 20 years ago, but that finding has not led to an effective therapy. The recent development stems from researchers' observation that the mutation causes mild or no symptoms in people who naturally produce fetal hemoglobin—a form normally produced only in the womb.

Hydroxyurea, the drug under study, has been used so far to treat about 60 patients with full-blown sickle cell anemia, according to study coauthor George J. Dover, a pediatrician at Johns Hopkins University School of Medicine. Among the 23 patients for whom the researchers have compiled complete data, the drug induced five to produce fetal hemoglobin at levels representing more than 20 percent of their total hemoglobin. In eight patients, fetal hemoglobin rose to a level of more than 10 percent. It represented less than 10 percent of total hemoglobin in five patients, while another five showed no response. At about the 20 percent level, disease symptoms begin to lessen, Dover says. The researchers do not understand fully how the drug works, but it appears to activate a gene controlling fetal hemoglobin production, he says.

Now hydroxyurea is undergoing a multicenter, 38-patient trial to determine maximum dose levels and the magnitude of the fetal hemoglobin response. But researchers must show more than an increase in fetal hemoglobin, Dover says. They need to demonstrate that this treatment improves patients' lives, which are interrupted by frequent hospital visits for painful episodes and infections resulting from blocked blood flow to vital organs. So far, it does not seem to produce any serious adverse effects, says Dover, who hopes to begin a controlled clinical trial of hydroxyurea in 1990.

Genetic evidence for autism

A surprisingly large percentage of parents and siblings of autistic individuals display language and personality features reminiscent of autism, suggesting a genetic component for this early-onset developmental disorder, reports Susan E. Folstein, a psychiatrist at Johns Hopkins.

In 2½-hour interviews, Folstein and her colleagues tested the general mental ability, social skills and language skills of normal parents and siblings of 40 autistic patients and a control group of 20 matched families of children with Down's syndrome. They concluded that about 30 percent of the family members of autistic subjects suffered pronounced reading and spelling difficulties as children and/or displayed language deficits as adults, particularly in social communication. They tended to be disorganized and overly detailed when speaking, Folstein says. In contrast, none of the controls showed pronounced deficits in these areas, she reports.

Autism, which appears in four out of every 10,000 children worldwide, is characterized by late speech development, repetitive or stereotyped behavior and an inability to develop social relationships, Folstein notes.

She says her findings and previous studies indicate the behavior of parents or siblings is not the cause of autism. Instead, she suggests the disease might arise from a recessive genetic defect. If two parents carry the relevant genes—leading to slight abnormalities in those individuals—then their child could inherit a more deleterious genetic combination resulting in autism, Folstein hypothesizes.

The ongoing study, which includes a research team in London, will eventually involve 240 "autistic" and 80 control families, she told SCIENCE NEWS.

Food Science

Egging on cholesterol-wary diners

Many people, concerned about their consumption of cholesterol-rich foods, have been weaning themselves from the egg. Now, hoping to revive sagging egg sales, the American Egg Board is crowing about new data showing U.S. eggs contain 23 percent less cholesterol than previously believed—about 213 milligrams per large egg.

The figure comes from a new nutrient assay directed by Gary R. Beecher, head of the Agricultural Research Service's Nutrient Composition Laboratory in Beltsville, Md. Previous "established" cholesterol values for eggs, published in 1976, were derived from journal articles, some published in the 1960s, Beecher notes. While he does not know the precise methods used to derive these data, Beecher says they invariably relied on techniques less specific to cholesterol than those his lab now uses. It's not that the old methods didn't measure cholesterol accurately, he says. "It's just that they measured a few other things as well—which may explain why their values were somewhat higher than ours."

His new data also show that previous nutrient values for iron, fat, vitamin B-12 and pantothenic acid (a B vitamin) overestimate levels in today's eggs by at least 10 percent. Beecher's data were gleaned from about 2,500 eggs collected from more than 100 farms—together responsible for handling 67 percent of marketed U.S. eggs.

Sweet solution to tainted poultry

Sweetening the drinking water of broiler chickens with lactose—or milk sugar—dramatically increases their ability to fend off infection by *Salmonella typhimurium*, a new study shows. Researchers estimate that this bacterium, which produces few if any symptoms in infected chickens, taints 5 to 30 percent of the 5.5 billion broiler chickens raised annually in the United States. It also appears responsible for roughly one-quarter of U.S. *Salmonella* food poisonings.

At the Agriculture Department's Veterinary Toxicology Research Unit in College Station, Tex., John R. DeLoach and his co-workers have been inoculating 3-day-old broiler chicks with 100 million *S. typhimurium* bacteria apiece. One week later, they inspect the birds for signs of infection. So far, all birds raised on regular drinking water have become heavily infected but only 53 percent of those given water containing 2.5 percent lactose show signs of the microbe. Moreover, untreated birds harbor 99.9 percent more bacteria than the lactose-treated birds that became infected. An interim report on the first three double-blind lactose trials will appear this summer in AVIAN DISEASES.

While DeLoach is still working to identify the mechanism behind the sugar's effect, his preliminary data do offer some hints. Lactose reduces the pH of a chicken's gut. DeLoach suspects the anaerobic bacteria fermenting this sugar produce short-chain fatty acids that can poison the *Salmonella*—provided the bird's small intestine has been sufficiently acidified. Only one other sugar—D-mannose, derived from a Mediterranean plant—appears comparable to lactose in effect, but it costs roughly 20 times as much.

Beet growers can breathe easy

Sugar beets continue to breathe after they're picked, using their stored sugars to fuel this cellular respiration. In fact, an estimated 250,000 tons of U.S. beet sugar is lost annually from crops awaiting processing. But a new strain of "lightly breathing" sugar beet, developed at the Agriculture Department's Northern Crop Science Lab in Fargo, N.D., is now available for crossing with existing commercial varieties. Not only does it hold the potential to cut breathing losses by 25 to 30 percent, but it's also resistant to the three most prevalent fungi responsible for storage rot.