

concerns about individual tolerance and adherence and about cost effectiveness at a population level. We think that in this situation, it is wise to use well-known drugs and, if possible, the less expensive ones. Enfuvirtide has no place in this setting in 2003.

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1. Ferranti S, Menichetti F. Enfuvirtide for prophylaxis against HIV infection. *N Engl J Med* 2003;349:815.
2. Beaman MH, Luft BJ, Remington JS. Prophylaxis for toxoplasmosis in AIDS. *Ann Intern Med* 1992;117:163-4.
3. Lazzarin A, Clotet B, Cooper D, et al. Efficacy of enfuvirtide in patients infected with drug-resistant HIV-1 in Europe and Australia. *N Engl J Med* 2003;348:2186-95.
4. Lalezari JP, Henry K, O'Hearn M, et al. Enfuvirtide, an HIV-1 fusion inhibitor, for drug-resistant HIV infection in North and South America. *N Engl J Med* 2003;348:2175-85. [Erratum, *N Engl J Med* 2003;349:1100.]

THE AUTHORS REPLY: Drs. Cuzin and Alvarez report that "in cases of high-risk exposure" the highest rate of HIV transmission is around 0.3 percent. Prospective studies suggest that this rate represents the av-

erage risk of transmission after percutaneous exposure to HIV-infected blood, but the risk is probably higher after exposure involving an increased volume of blood or a high viral load,^{1,2} both of which might apply to health care workers exposed to patients with resistance to antiretroviral drugs — a possible reason for the failure of postexposure prophylaxis. We recommend the restricted use of prophylactic enfuvirtide in high-risk persons,³ such as health care workers, in a setting where the storage, preparation, and administration of the drug are performed by skilled personnel.

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1. Updated U.S. Public Health Service guidelines for the management of occupational exposures to HBV, HCV, and HIV and recommendations for postexposure prophylaxis. *MMWR Recomm Rep* 2001;50(RR-11):1-52.
2. Bell DM. Occupational risk of human immunodeficiency virus infection in healthcare workers: an overview. *Am J Med* 1997;102: Suppl 5B:9-15.
3. Ferranti S, Menichetti F. Enfuvirtide for prophylaxis against HIV infection. *N Engl J Med* 2003;349:815.

Genomic Medicine

TO THE EDITOR: In their editorial (Sept. 4 issue),¹ Guttmacher and Collins provide a conventional and optimistic overview of the effects of genomics on clinical practice. Absent from their discussion is mention of the most common current use of genetics in clinical practice: negative eugenics. Testing for chromosomal disorders is routine in obstetrics and fertility medicine. The example of Tay–Sachs disease demonstrates that voluntary screening of the population and genetic counseling can have a substantial effect on the incidence of disease.² As the novelist Josef Skvorecký has written, "the past shows the potential of the future."³ Increasing knowledge of genetic contributions to disease increases opportunities for eugenic intervention. This is not a theoretical possibility; the organizations that sponsored screening for Tay–Sachs disease are expanding this program to include other genetic diseases that are relatively common among Ashkenazi Jews.⁴ To date, translation of the identification of the gene for a disease into effective therapy has been uncommon. Some disorders with substantial genetic components such as schizophrenia and autism probably

result from developmental processes that are difficult to manipulate, and eugenic intervention will be easier than developing medical or gene therapy. The genomic era will also be an era of eugenics.

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1. Guttmacher AE, Collins FS. Welcome to the genomic era. *N Engl J Med* 2003;349:996-8.
2. Kaback MM. Population based genetic screening for reproductive counseling: the Tay Sachs experience. *Eur J Peds* 2000;159: S192-S195.
3. Skvorecký J. *The miracle game*. New York: Alfred A. Knopf, 1991.
4. Kolata G. Using genetic tests, Ashkenazi Jews vanquish a disease. *New York Times*. February 18, 2003:F1.

DRS. GUTTMACHER AND COLLINS REPLY: Dr. Albin's letter raises an important issue: to what extent will the discovery of susceptibility genes for common disorders such as cancer, diabetes, obesity, autism, and schizophrenia lead to their use in the prenatal arena, resulting in the termination of pregnancies? Three factors militate against this practice's becom-

ing widespread: the predictive value of genetic testing for non-mendelian conditions will remain poor, even when all the hereditary factors have been identified; public interest in using prenatal diagnosis and the termination of pregnancy for disorders with less profound implications and less precise predictability than those associated with Tay–Sachs disease will most likely be rather limited¹; and knowledge of the relevant genetic pathways is expected to lead to more effective therapeutic interventions in the future.

We share a concern, however, about a related circumstance in which the widespread availability of preimplantation genetic diagnosis might shift the balance toward “opting in” for “desirable” embryos, as opposed to “opting out” through the ter-

mination of pregnancy. This topic is but one of many being studied by the Ethical, Legal, and Social Implications program of the National Human Genome Research Institute.² Ultimately, the broad community will have to be the judge of the wisdom and ethics of such applications.

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1. Lodder AE, Frets PG, Trijsburg RW, Meijers-Heijboer EJ, Klijn JGM, Niermeijer MF. Attitudes towards termination of pregnancy in subjects who underwent presymptomatic testing for the BRCA1/BRCA2 gene mutation in the Netherlands. *J Med Genet* 2000;37:883-4.

2. Collins FS, Green ED, Guttmacher AE, Guyer MS. A vision for the future of genomics research. *Nature* 2003;422:835-47.

Case 27-2003: A 36-Year-Old Man with Recurrent Epigastric Pain and Elevated Amylase Levels

TO THE EDITOR: In Case 27-2003 (Aug. 28 issue),¹ a patient with two rare diseases — lymphoplasmacytic sclerosing pancreatitis and amyotrophic lateral sclerosis — is described. The pathological discussion concludes with the statement that these two diseases are unrelated, but as William Osler emphasized more than a century ago, the astute clinician should attempt to find a single explanation for all the symptoms. Could both illnesses have had a single underlying cause? Lymphoplasmacytic sclerosing pancreatitis has been described as autoimmune pancreatitis, and it has been suggested that autoimmunity is a cause of amyotrophic lateral sclerosis.² Viruses can cause pancreatitis and are a possible cause of amyotrophic lateral sclerosis.³ It should be determined whether this unusual combination of illnesses has been diagnosed in other patients.

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1. Case Records of the Massachusetts General Hospital (Case 27-2003). *N Engl J Med* 2003;349:893-901.

2. Appel SH, Smith RG, Alexianu MF, Engelhardt JI, Stefani E. Autoimmunity as an etiological factor in sporadic amyotrophic lateral sclerosis. *Adv Neurol* 1995;68:47-57.

3. Rowland LP, Shneider NA. Amyotrophic lateral sclerosis. *N Engl J Med* 2001;344:1688-700.

TO THE EDITOR: I read with interest the recent case of a young man with probable autoimmune pancre-

atitis. I am an avid follower of the Case Records and am especially fond of the discussion building up to the diagnostic tests. I was surprised to read, however, that a Whipple procedure was considered diagnostic. If partial abdominal evisceration is thought to be diagnostic, then what procedure would be considered therapeutic?

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THE DISCUSSANT REPLIES: Dr. Lowenfels is correct in pointing out the potential association between the two rare diseases affecting this patient. Current data indicate that lymphoplasmacytic sclerosing pancreatitis is an autoimmune disorder, and it has been linked to many other autoimmune diseases. Since the cause of amyotrophic lateral sclerosis is not known, and since autoimmunity may be an etiologic factor, one can certainly speculate that the two diseases are related. This association may be strengthened if similar cases are reported.

Dr. Makins remarks on the fact that the Whipple procedure was performed for diagnostic purposes. Perhaps it would have been more appropriate to label the operation a diagnostic–therapeutic procedure. Although lymphoplasmacytic sclerosing pancreatitis was suspected (and although the patient actually received treatment with corticosteroids for