

father's cells showed decreased O₂ uptake below the average for normal males (P/B of 9 compared to normals of 11.83 ± 1.94).

It is suggested from these studies that leukocyte O₂ uptake may be of value in detecting asymptomatic carriers of CGD for more precise genetic counseling.

α -1,4 Glucosidase activity in Pompe's disease. IRA S. SALAFSKY and HENRY L. NADLER. *Northwestern Univ. Med. Sch., Children's Mem. Hosp., Chicago, Ill.*

α -1,4 Glucosidase (α -glu) has been shown to be absent in cultivated amniotic fluid cells (AFC), fibroblasts (FIB) and liver (LIV) of patients with Pompe's disease. In contrast, α -glu is present in the kidney (KID) and amniotic fluid (AF) of these patients. In an attempt to explain these differences, properties of α -glu including pH optima, turanose inhibition and heat inactivation were studied in controls (C) and Pompe's (P). The results are as follows:

	Specific Activity*		pH Optima		Turanose Inhibition†		Heat Inactivation‡	
	C	P	C	P	C	P	C	P
AF	5.4	14	6.0	6.0	47	51	7	8
AFC	5.1	0	4.0	—	92	—	100	—
FIB	5.9	0	4.0	—	90	—	100	—
LIV	9.3	0	4.0	—	94	—	100	—
KID	10.3	7.5	6.0	6.0	33	53	100	100

* umoles maltose hydrolyzed/min/gm protein.

† % inhibition by 0.03 M turanose.

‡ % activity remaining after heating at 45°C for 15 minutes.

No α -glu activity could be demonstrated in urine or maternal serum. α -glu in amnion had properties identical with FIB and LIV enzyme. Upon differential centrifugation of the cell-free amniotic fluid, cells and tissues, α -glu was found primarily in the 25,000 × g fraction. These data clearly indicate that the α -glu present in AFC, FIB and LIV have identical properties which are distinctly different from the α -glu in AF and KID. The α -glu in AF differs from that found in KID in its greater heat lability. These studies fail to identify the origin of the α -glu in amniotic fluid. The diagnosis of Pompe's disease *in utero* must rest on the demonstration of the α -glu deficiency in amniotic fluid cells.

Testicular feminization: Expression in sex skin fibroblast culture.

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The inheritance and pathogenesis of human testicular feminization (TF) are unknown. The possibilities of X-linked recessive inheritance and primary target organ refractoriness to testosterone (T) could be studied optimally in serially subcultured cell strains. Fibroblasts derived from sex and non-sex skin of males, females and patients with TF were incubated with T-4-¹⁴C. T and its metabolites in the culture medium were separated by paper chromatography and identified by reverse isotopic dilution. The rate of T metabolism was much greater in sex than in non-sex strains. For both newborn foreskin and adult labial strains, androstenedione was the primary metabolite to T, however they differed in their rate of production of 5 α -androstenedione and andosterone. In a strain derived from the foreskin of a 7-year-old, 5 α -androstenedione, rather than androstenedione, appeared to be the major early metabolite. 5 α -dihydro-testosterone (DHT) was

a major metabolite in none of the strains. Sex and non-sex strains from two patients with TF were indistinguishable: Both had low rates of T metabolism. We conclude that: 1) *in situ* differences in T metabolism between normal sex and non-sex skin persist in their serially subcultured fibroblasts; 2) an expression of the TF gene is detectable in sex skin fibroblasts without reference to the rate of DHT formation; and 3) Lyonization of cloned heterozygous fibroblasts may prove that human TF is X-linked.

Evidence linking an extra Y chromosome to sociopathic behavior. LYTT I. GARDNER and RICHARD L. NEU. *State Univ. of New York Upstate Med. Ctr., Syracuse, N. Y.*

There is accumulating evidence that certain XYY males are more prone to sociopathic behavior than are XY males, and that the extra Y chromosome may be causally related to this behavior. Eleven studies were surveyed which the percentage of XYY were identified among selected male populations in prisons and mental hospitals. The men were selected for height (over 59 in.), having dangerous or violent propensities, or for being mentally subnormal. These percentages were contrasted with the findings of XYY males in several surveys of newborn populations. There was found a strikingly higher incidence (1.8 to 12.0 per cent) of XYY males in institutions than in the general populations as determined from newborn surveys (0.14 to 0.38 per cent). It could appear that only a small fraction of the total numbers of XYY males known to exist in the population are institutionalized sociopaths. The XYY sociopaths represent a numerically small subdivision of the large group of mostly normal XYY individuals (there is some indication that a nosologic classification is developing, since XYY males with abnormal genitalia appear to represent a discrete subgroup). The implications of these data are obvious in the counselling of parents of XYY children. It is especially important that the physician allay the fears created in parents by the numerous popular articles linking the XYY karyotype to criminality through a careful presentation of the data thus far at hand.

Perinatal expression of the Lewis and secretor blood group systems. MINERVA B. ARCILLA and PHILLIP STURGEON. *U. C. L. A. Sch. of Med., Los Angeles, Calif.*

Studies on expression at birth and on maturation of the secretor and Lewis blood group systems, as manifest on red cells and in saliva, have been carried out on a series of infants using three Lewis reagents: anti-Le^a, -Le^b and -Le^x.

Among adults, 20% are Lewis red cell type Le(a+b-x+) and 70% are Le(a-b+x+); both types are essentially absent at birth. A third type Le(a-b-x-) is found in both 10% of adults and in 10% of infants; 90% of newborn infants, however, have the unique type Le(a-b-x+). Infants of the latter type undergo a rapid maturation during the first week of life to the adult Le(a+b-x+) type. Then, if they are salivary non secretors of ABH, they remain that type throughout life. Whereas those who are secretors evolve during the ensuing four months to Le(a+b+x+) which is also a type practically unique to infancy; finally, by two years of age, they evolve to the most common adult type, Le(a-b+x+).

Salivary phenotypic expression of the genes involved in the above red cell types is found in soluble form but the state of development at birth is more advanced than on the red cell and the subsequent maturation is more rapid. Prematures show a relatively delayed maturation in the expression of these genes. Thus, unlike other blood group systems, the Lewis system is unique in infancy; it shows differential tissue expressivity and,