Biomedical update:

Tourette's: one in every 100 boys?

One in 100 boys has some form of Tourette Syndrome, according to a surprising study of 3,304 elementary school students in southern California. Geneticist David E. Comings also reports, in the *Journal of Clinical Psychiatry*, that one in 759 girls shows signs of the disorder. Comings' figures are far higher than previous estimates of the disorder.

In addition, Comings and colleagues found that about 30 percent of relatives of individuals with Tourette syndrome have some form of behavior disorder, compared with six percent of relatives of control group subjects. Problems seen in relatives ranged from anxiety, depression, and obsessive-compulsive behavior to drug and alcohol abuse, eating disorders, sexual disorders, and violent behaviors.

The symptoms of Tourette syndrome itself include tics such as eyeblinking or lip smacking, and uncontrolled utterances such as grunting, coughing, or cursing. People with Tourette syndrome also may exhibit compulsive behaviors, dyslexia or other learning disabilities, short temper, depression, phobias, and anxiety attacks. Tourette syndrome often co-occurs with autism, leading some researchers to believe the two disorders are related—a connection which may be harder to prove, if Comings' high general-population figures are verified.

Comings believes Tourette syndrome may be caused by the inheritance, from both parents, of a gene limiting the availability of the brain "messenger" chemical serotonin. A shortage of serotonin could interfere with the transmission of messages between the limbic system (which governs emotions and behavioral responses to stimuli) and the frontal lobes of the brain, which are involved in planning and judgment.

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"The ticcing link," Bruce Bower, *Science News*, Vol. 138, July 21, 1990; pp. 42-44.

Autism, defect on chromosome 15 reported in one case

There is evidence that a defective gene or genes on a segment of chromosome 15 is linked to Prader-Willi syndrome (which causes retardation and extreme obesity due to compulsive eating) and "happy puppet" syndrome (which causes puppet-like movements and frequent abnormal bursts of laughter). Now Jacob Kerbeshian et al. report the case history of a woman with a deletion of a chromosome 15 segment (q12), autism, profound mental retardation, and manic depression.

Kerbeshian's 33-year-old subject also had unusual facial features reminiscent of fetal alcohol syndrome (although there was no evidence of maternal alcohol abuse), and frequently attacked others violently. Like a number of other autistic individuals with manic depressive disorder, she responded positively to treatment with lithium.

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"Autism, profound mental retardation, and atypical bipolar disorder in a 33-year-old female with a deletion of 15q12," J. Kerbeshian, L. Burd, T. Randall, J. Martsolf, and S. Jalal; *Journal of Mental Deficiency Research*, 1990, 34, pp. 205-210. Address: Larry Burd, Medical Center Rehabilitation Hospital, 1300 South Columbia Road, Grand Forks, North Dakota 58202.

Males with Rett reported

Rett syndrome, a "new" disorder identified in 1966 by Andreas Rett, remains a frustrating puzzle, with exhaustive studies offering few clues about its cause. Now, even the one generally accepted assumption about Rett—that it affects only females—is being questioned.

Last year, ARRI reported that Michel Philippart of UCLA had informally mentioned seeing several adult males who appeared to have Rett syndrome. Philippart has now published a paper describing the men, whose symptoms and histories he says are "indistinguishable from typical females with the Rett syndrome."

Philippart's subjects both developed normally, and then developed symptoms of Rett syndrome including unusual hand movements and loss of purposeful hand use, seizures, mental and motor deterioration, scoliosis, and loss of previously learned language. Unlike many individuals with Rett, the men did not show significant autistic behaviors at any stage, although one man's yelling, moaning and odd movements did resemble autistic behavior.

It makes sense, Philippart, that Rett syndrome occurs in men as well as women. "Since there are no known sexual differences in brain structure or developmental stages," he says, "any syndrome observed in females should also be observed in males." It is "utterly unclear," he says, why Rett is so rarely seen in men.

The early stages of Rett syndrome are often mistaken for autism, because children may become withdrawn, lose language skills, and develop symptoms which resemble autistic self-stimulation. Later, autistic behaviors tend to disappear but physical and mental deterioration accelerate.

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"The Rett syndrome in males," Michel Philippart; *Brain and Development*, Vol. 12, No. 1, 1990, pp. 33-36. Address: Michel Philippart, Neuropsychiatric Institute, UCLA, 760 Westwood Plaza, Los Angeles, CA 90024.

Elbow splints for Rett's

The constant hand-wringing, handwashing, clapping, and hand-to-mouth movements seen in Rett Syndrome—clinical features so striking and universal that the logo of the International Rett Syndrome Association is two clasped hands—can be reduced by the use of plastic elbow splints, occupational therapist Marilyn Aron reports.

Aron notes that girls with Rett often bite their hands or wrists, which can lead to infection, skin irritation around the mouth, or blistering, cracking, and bleeding of the hands.

Aron had splints designed from Polyform™ plastic with Polycushion™ linings that do not absorb odors or encourage bacteria. She tested the splints on eight girls with Rett syndrome whose hand movements caused self-injury, and reports that splinting appeared to have several beneficial effects: hand-to-mouth movements were inhibited, hand-wringing also decreased, several girls appeared calmer, and all parents reported increased socialization. Teachers and therapists also reported that the girls improved in such areas as grasping, using walkers, and understanding of objects around them.

"The splints should not be viewed as restraints," Aron says, "but rather as an adjunct to treatment."

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"The use and effectiveness of elbow splints in Rett Syndrome," Marilyn Aron, in press. Address: Marilyn Aron, 600 Scenic Drive, Ashland, OR 97520.

DPT claim deadline near

The federal vaccine injury compensation system established under the National Childhood Vaccine Injury Compensation Act of 1986 has paid $31.5 million for 60 victims injured or killed by vaccines. The awards are the first to be paid under a law signed by President Reagan in 1986 after five years of effort by Dissatisfied Parents Together (DPT), the American Academy of Pediatrics, and vaccine manufacturers to create the nation's first federal vaccine injury compensation system. The deadline for filing a claim for individuals killed or injured before Oct. 1, 1988, is Sept. 30, 1990.

Compensation is paid for injuries or deaths resulting from mandated childhood vaccines, including diphtheria, pertussis, tetanus, measles, mumps, rubella, and polio. Vaccine injuries may range from minor brain damage to severe and profound mental retardation, deafness, blindness, loss of motor function and seizures. Qualification is based on medical records and a table of compensable events built into the law. For more information call the National Vaccine Information Center, (703) 938-DPT3, or U.S. Claims Court, (202) 633-7257.