

heritable marks established in the germ line according to the parental origin of alleles. This is the process of gene imprinting, wherein only one allele of a gene is expressed in somatic cells. Technically, however, the epigenetic process we review

with respect to cancer cells is, as Dr. Kirk points out, a somatic-cell transmission mechanism.

James G. Herman, M.D.

Stephen B. Baylin, M.D.

Johns Hopkins Medical Institutions

Baltimore, MD 21231

sbaylin@jhmi.edu

## Congenital Pulmonary Lymphangiectasia

**TO THE EDITOR:** In Hagmann and Berger's description of congenital pulmonary lymphangiectasia in *Images in Clinical Medicine* (Nov. 27 issue),<sup>1</sup> the authors state, "Congenital pulmonary lymphangiectasia . . . is a uniformly fatal disease when it manifests in the newborn period." Having followed several such patients for 12 to 60 months, we disagree with this statement. The authors evidently did not see the report on radiographic findings in children with congenital pulmonary lymphangiectasia who survive infancy.<sup>2</sup> Much of the literature on congenital pulmonary lymphangiectasia is heavily influenced by early reports that preceded modern care provided in the neonatal intensive care unit. We believe that patients with suspected congenital pulmonary lymphangiectasia should be treated aggressively, since at least a subgroup of them have a good prognosis for long-term survival.

Jonathan Finder, M.D.

Jonathan Steinfeld, M.D.

Children's Hospital of Pittsburgh

Pittsburgh, PA 15213

finder@pitt.edu

1. Hagmann C, Berger TM. Congenital pulmonary lymphangiectasia. *N Engl J Med* 2003;349:e21 (Web only). (Available at [www.nejm.org/cgi/content/full/349/22/e21](http://www.nejm.org/cgi/content/full/349/22/e21).)

2. Chung CJ, Fordham LA, Barker P, Cooper LL. Children with congenital pulmonary lymphangiectasia: after infancy. *AJR Am J Roentgenol* 1999;173:1583-8.

nary lymphangiectasia associated with pulmonary venous obstruction, and congenital pulmonary lymphangiectasia associated with a generalized defect in lymphatic development. Pulmonary involvement is most severe in patients with isolated congenital pulmonary lymphangiectasia, and the prognosis for the majority of these patients remains poor even with modern care provided in a neonatal intensive care unit.<sup>2,3</sup> However, there are some reports of long-term survivors.<sup>3-5</sup> Although not clearly specified in the report by Chung et al.,<sup>4</sup> the onset of symptoms was delayed beyond the neonatal period in the three survivors with isolated congenital pulmonary lymphangiectasia who were described by Bouchard and colleagues.<sup>3</sup> Therefore, as Drs. Finder and Steinfeld correctly point out, isolated congenital pulmonary lymphangiectasia may indeed not be a uniformly fatal disease; its prognosis appears to depend on the severity of symptoms observed in the immediate postnatal period.

Thomas M. Berger, M.D.

Cornelia Hagmann, M.D.

Children's Hospital of Lucerne

CH-6000 Lucerne, Switzerland

thomas.berger@ksl.ch

Jan-Olaf Gebbers, M.D.

Kantonsspital Luzern

CH-6000 Lucerne, Switzerland

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1. Noonan JA, Walters LR, Reeves JT. Congenital pulmonary lymphangiectasias. *Am J Dis Child* 1970;120:314-9.

2. Case Records of the Massachusetts General Hospital (Case 13-1992). *N Engl J Med* 1992;326:875-84.

3. Bouchard S, Di Lorenzo M, Youssef S, Simard P, Lapierre JG. Pulmonary lymphangiectasia revisited. *J Pediatr Surg* 2000;35:796-800.

4. Chung CJ, Fordham LA, Barker P, Cooper LL. Children with congenital pulmonary lymphangiectasia. *AJR Am J Roentgenol* 1999;173:1583-8.

5. Scott C, Wallis C, Dinwiddie R, Owens C, Coren M. Primary pulmonary lymphangiectasia in a premature infant: resolution following intensive care. *Pediatr Pulmonol* 2003;35:405-6.