EDITORIAL

PLASTIC BRONCHITIS, CHYLOUS REFLUX, AND LYMPHATIC IMAGING: A CONTINUING STORY

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Keywords: plastic bronchitis, chyloptysis, chylous reflux, LAM, lymphatic imaging, Herbert Maier

In 1973, during the 4th International Congress on Lymphology in Tucson, thoracic surgeon Herbert Maier, a devout card-carrying lymphologist, chatted in our office about his latest mission – to raise awareness of what he coined “chyloptysis” and the neglected role of the “(cardio)thoracic lymphatic system” in a variety of rare and common lymphatic disorders. As a daily reminder of the body’s vulnerability to dysfunction of the interstitial fluid drainage system, he himself had suffered from a recurrent craniopharyngioma for more than 20 years plagued by a continuous nasal leak of cerebrospinal fluid through his re-operated and chronically infected cribiform plate. In a visionary yet long buried paper (1) from his 1966 Presidential Address to the American Thoracic Society, he called attention to the lymphatics of the chest as the next frontier for thoracic surgeons. Further, he confessed to us his frustrating encounters with a little known usually fatal lung disease without a name (now called lymphangioleiomyomatosis or LAM), which he viewed as a prime example of disordered pulmonary lymphatic proliferation, influenced by female hormones, and commonly manifesting as chylous effusions and occasionally as “chyloptysis” – expectoration of milky intestinal lymph. He cared compassionately and alone for a group of these LAM patients and improved their outlook with a variety of operative approaches to lymphatic complications, rerouting the lymph circulation, and suppressing hormonal influences. At the same time, the newly formed International Society of Lymphology (ISL) in its earliest Congresses (2) was also focusing on improved visualization of the previously inaccessible peripheral and central lymphatic system by invasive lipid-soluble contrast lymphography (LAG) (Kinmonth, Viamonte, Ruttimann, Wallace, Kandl) and also featuring visceral chylous reflux syndromes and their treatment including dietary and operative approaches [surgeons from Belgium (Godart, Gruwez), France (Servelle), Italy (Tossati) and Brazil (Degni, Cordeiro) among others].

In this issue of Lymphology, in an era where cardiac surgery has reached new heights in rescuing infants born with previously unsurvivable complex congenital cardiac anomalies, Herbert Maier’s simple and timeless observations resonate with the case report of a child successfully treated for life-threatening “plastic bronchitis” manifesting as chyloptysis after a series of cardiovascular operations leading up to a Fontan procedure for hypoplastic left ventricle (3). Indeed, the Pediatric Cardiology and Pulmonary Divisions at the University of
Arizona have seen three such cases arising years after a successful Fontan procedure, with two of the children surviving long-term and now leading near normal lives. In part this success was due not only to careful control of the cardiovascular system and hemodynamics, i.e., dictated by the status of the heart and blood vessels (viz, arteries, veins, and blood capillaries) but especially from particular attention to the lymphodynamics and that other vasculature – the lymphatic system. Dynamic minimally invasive bipedal whole body lymphatic imaging (lymphangioscintigraphy) precisely pinpointed the area of lymph reflux into the tracheobronchial tree and thereby the site of formation of near daily suffocating chylous bronchial casts. Initially, octreotide slowed intestinal lymph flow and subsequently, simple dietary manipulation eliminating dietary fat and the long-chain triglycerides that form chylomicra, sufficed to control the condition for several years. Maintenance therapy with captopril further reduced cardiac afterload (and central venous pressure) – likely decreasing lymph formation and promoting lymph return to the central venous system (4) – and subsequently, thoracic duct ligation, when dietary compliance became an issue several years later, completed the formula for clinical success and improved quality of life. It remains unknown whether the lymphatic disorder producing chyloptysis so prominently after the Fontan procedure was entirely secondary to operative interruption, pressure-flow disturbances, or complicated by associated congenital lymphatic abnormalities. Indeed, cardiac anomalies not uncommonly are featured in a variety of congenital including hereditary lymphedema (“lymphologic”) syndromes (5), particularly those involving disturbed valve formation common to the heart, veins, and lymphatics. Indeed, multiple genes and corresponding proteins have been specifically implicated in this process and underlie human disorders and transgenic mouse models manifesting lymphedema and/or chylous effusions as well as cardiac and venous anomalies (6).

Nonetheless, this case report and the three others that follow it in this issue of Lymphology, each an “n-of-1 clinical trial” (7), reemphasize the importance of the lymphatic system in complex, rare but now treatable cardiopulmonary and abdominal visceral disorders as well as in common conditions such as portal hypertension/ hepatic cirrhosis and congestive heart failure (4,8). Importantly, lymphatic system pathophysiology can be vividly and routinely displayed by bipedal lymphangioscintigraphy (even in the neonate), and supplemented by indocyanine green (ICG) and magnetic resonance lymphography although conventional oil contrast LAG may be necessary for fine delineations prior to lymphatic surgery. Further, the success of relatively simple straightforward dietary and operative maneuvers can control the life-threatening lymph reflux (9).

In the 1950’s and 1960’s, leading up to the founding of the International Society of Lymphology nearly 50 years ago, visualization of a wide variety of human lymphatic diseases through invasive conventional LAG and documentation of the lymphodynamics of common disturbances such as hepatic cirrhosis and congestive heart failure (4,8) by thoracic duct cannulation and drainage drew attention to the broad clinical relevance of the lymphatic system. Now in the era of molecular lymphology, advanced multimodal lymphatic imaging, and spectacular cardiac and microvascular surgical advances, thoracic surgeon-lymphologist Herbert Maier’s prescient challenge remains to further explore lymphatic system physiology, pathophysiology, and interrelationships with other body systems, dissect molecular mechanisms, and translate these insights into new and effective non-operative and operative approaches. In the final analysis, the lymphatics may be at the heart of these rare and many other perplexing common conditions.
REFERENCES


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