Lipedema and Lipolymphedema

Lipedema, initially described at the Mayo clinic in 1940,[1] [2] is a loose, connective-tissue (fat) disease (lipomatosis) with a pathological deposition of fibrotic fatty tissue on the limbs of women sparing the trunk, hands and feet,[3] [4] resulting in a disproportionate body habitus. There is no specific ICD-10-CM code for lipedema. Deposition of lipedema fat increases with stage and body mass index (BMI) and likely involves sex hormones during times when weight is gained (puberty, pregnancy and menopause). Lipedema is inherited in 60% of women likely through genes affecting microvessels resulting in excess fluid bound to glycosaminoglycans in the interstitial space.[5]

Unique to lipedema is fat that is highly resistant to loss by diet, exercise, or bariatric surgery.[6] [7] [8] Lipedema is often confused with secondary obesity or lymphedema. Women with lipedema and/or obesity can develop lymphedema called lipolymphedema, for which there is no ICD-10-CM code.

There is no cure for lipedema, but treatments aimed at reducing the lymphedema component of lipedema such as manual decongestive therapy, wrapping, exercise, compression garments and pumps, and some medical foods and medications are helpful. Expertly performed suction assisted lipectomy is the treatment of choice for suitable lipedema patients with an inadequate response to conservative and supportive measures.[9]

Lipedema is thought to affect 11% of the female population,[10] with rates of 6-39% reported.[11] With a total US population of 330 million, and 50.8% female (census.gov), as many as 18 million women in the United States could have lipedema–and the majority have not been diagnosed.

Specific ICD-10-CM codes will improve identifying, tracking, and research on women with lipedema and help distinguish it from other lipomatosis diseases, including Madelung's disease, a rare disease of fat primarily on the upper body, and Dercum's disease, a rare disease whose hallmark is painful lipomas. Lipedema, Madelung's disease and Dercum's disease merit specific codes that capture their unique characteristics and distinguish them from one another. Multiple symmetric lipomatosis or Launois Bensaude lipomatosis, synonyms for Madelung's disease, are also included.[12]

The ICD-10 codes currently used in Germany have specific codes for lipedema and its three stages (E88.20-E88.22) and Dercum's disease; therefore, efforts to match these codes to facilitate research between countries is important.

ICD-10-CM Codes

Adopting specific ICD-10-CM codes for three stages of lipedema, and related but separate and distinct lipomatosis diseases, has a number of benefits, and the submitter believes this would aid: epidemiologic monitoring, assessment of disease-associated medical costs, retrospective studies comparing best practices, recruitment of participants for clinical trials and patient registries, improved assessment of resource requirements, and alignment with the ICD-10 codes in Germany.

Women with lipedema Stage 3 are very different than women with Stage 1 or 2, therefore separate codes for stages is important for prevalence studies, determination of progression or regression of stages and for clinical assessment and care. Descriptions of the three stages of lipedema are as follows:

Stage 1: Normal skin surface with enlarged hypodermis (lipedema fat).

Stage 2: Uneven skin with indentations in fat and larger hypodermal masses.

Stage 3: Bulky extrusions of skin and fat causing large deformations especially on the thighs and around the knees that drastically inhibit mobility.[13] [14]

In 2020 lipedema is coded using several different ICD-10-CM codes: R60.9, Q82.0, and E82.0. Lipedema is only found in the ICD-10-CM index crosslinked to R60.9: Edema. None of these codes are specific or adequate.

Included in code E82.0 are adiposis dolorosa, Dercum's disease, and lipomatosis. Adiposis dolorosa is an outdated term used incorrectly to describe lipedema and Dercum's disease and we are requesting removing the term from ICD-10-CM.[15] People with Dercum's disease have painful lipomas in subcutaneous fat from scalp to

foot. Lipomatosis, Not Otherwise Specified, is a benign usually autosomal dominant condition in families with multiple lipomas on the body, most often the trunk and extremities.[16] These diseases are discrete conditions from lipedema and unique codes for each are proposed.

Lymphedema is a chronic and progressive swelling caused by a low output failure of the lymphatic system, resulting in the development of a high-protein edema in the tissues. Lymphedema is a lifelong condition for which no cure exists.[17] An estimated 250 million people are affected by lymphedema worldwide.[18] Lymphedema can be either primary (hereditary) or secondary. Secondary lymphedema is the most common cause of the disease and affects approximately 1 in 1000 Americans.[19] Complications of lymphedema include recurrent bouts of cellulitis and/or lymphangitis, bacterial and fungal infections, lymphangio-adenitis, deep venous thrombosis, poor wound healing, leg ulcers, severe functional impairment, disability, and necessary amputation. Patients with chronic lymphedema for 10 years have a 10% risk of developing lymphangiosarcoma.[20] The 5-year survival rate for lymphangiosarcoma is less than 10%.[21] Praecox lymphedema is currently misclassified in ICD-10-CM as a secondary lymphedema; it is more accurately classified under code Q82.0: Hereditary lymphedema.[22]

Lymphedema can occur secondary to obesity[23] [24] *or* lipedema, both forms of lipolymphedema secondary to a fat excess. Lymphedema is a common comorbid condition in severe obesity individuals with BMIs greater than 50 Kg/ m^2. In the US the prevalence of obesity was 42% in 2017-18 and is increasing.[25] Lipolymphedema is likely the second most common of lymphedema and may soon become the most common cause of lymphedema. Lipolymphedema, is not in the ICD-10-CM index.

The Lipedema ICD 10-CM Committee, with support from the American Vein & Lymphatic Society (AVLS) is submitting the following modifications to ICD-10-CM to identify and track lipedema and lipolymphedema patients.

TABULAR MODIFICATIONS

DELETE ~~E88.2 Lipomatosis, not elsewhere classified~~

DELETE ~~Adiposis dolorosa~~

ADD SUBCATEGORY

E88.2 Lipomatosis

ADD Code also *pain*

NEW CODE E88.20 Lipedema, Stage 1

NEW CODE E88.21 Lipedema, Stage 2

NEW CODE E88.22 Lipedema, Stage 3

NEW CODE E88.23 Lipedema, Not Otherwise Specified

NEW CODE E88.24 Dercum's Disease

NEW CODE E88.28 Madelung's disease

ADD Multiple symmetric lipomatosis

ADD Launois Bensaude lipomatosis

NEW CODE E88.29 Lipomatosis NOS

E88.89 Metabolic disorder, unspecified

DELETE ~~Madelung's disease~~

DELETE ~~symmetrical lipomas, neck~~

R60.9 Edema, unspecified

DELETE ~~lipedema~~

	I89.0	Lymphedema, not elsewhere classified
		Lymphedema (acquired)
		Elephantiasis (nonfilarial) NOS
		Lymphangiectasis
		Obliteratino, lymphatic vessel
DELETE		~~Praecox lymphedema~~
	Q82.0	Hereditary lymphedema
ADD		Praecox lymphedema
NEW CODE	I89.3	Lipolymphedema

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