&OLQLFDO SUR¿OH UHVSRQVH WR WKHUDS\ DQG RXWFRPH RI S

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Objective: Intestinal lymphangiectasia (IL, primary or secondary) is an important cause of protein-losing enteropathy. We evaluated the clinico-laboratory pro le, response to therapy, complications and outcome of childre with primary IL (PIL).

Methods:Consecutive children (18 years) diagnosed with PIL (clinical setting, typical small bowel histopathology and exclusion of secondary causes) from 2007 to 2017 were evaluated.

Results:28 children with PIL (16 boys, age at symptom onset-12 months and at diagnosis 8 years) were studie Pedal edema (93%), chronic diarrhea (78.6%) and recurrent anasarca (64%) were the common presentatio Ascites, pleural and pericardial e usion was seen in 64% (n-18; chylous-5, non-chylous-13), 18% and 18% ca respectively. Hypoproteinemia, hypoalbuminemia, hypocalcaemia and lymphopenia were present in 82%, 82 75% and 39% cases respectively. Duodenal biopsy established the diagnosis in 86% cases, while 14% req distal small bowel biopsies. Dietary therapy was given in all and 6 cases required additional therapy (octreotide tranexamic acid-3 and total parenteral nutrition-1). Lymphedema (3/5 vs. 1/23), pleural e usion (4/5 vs. 1/23) and