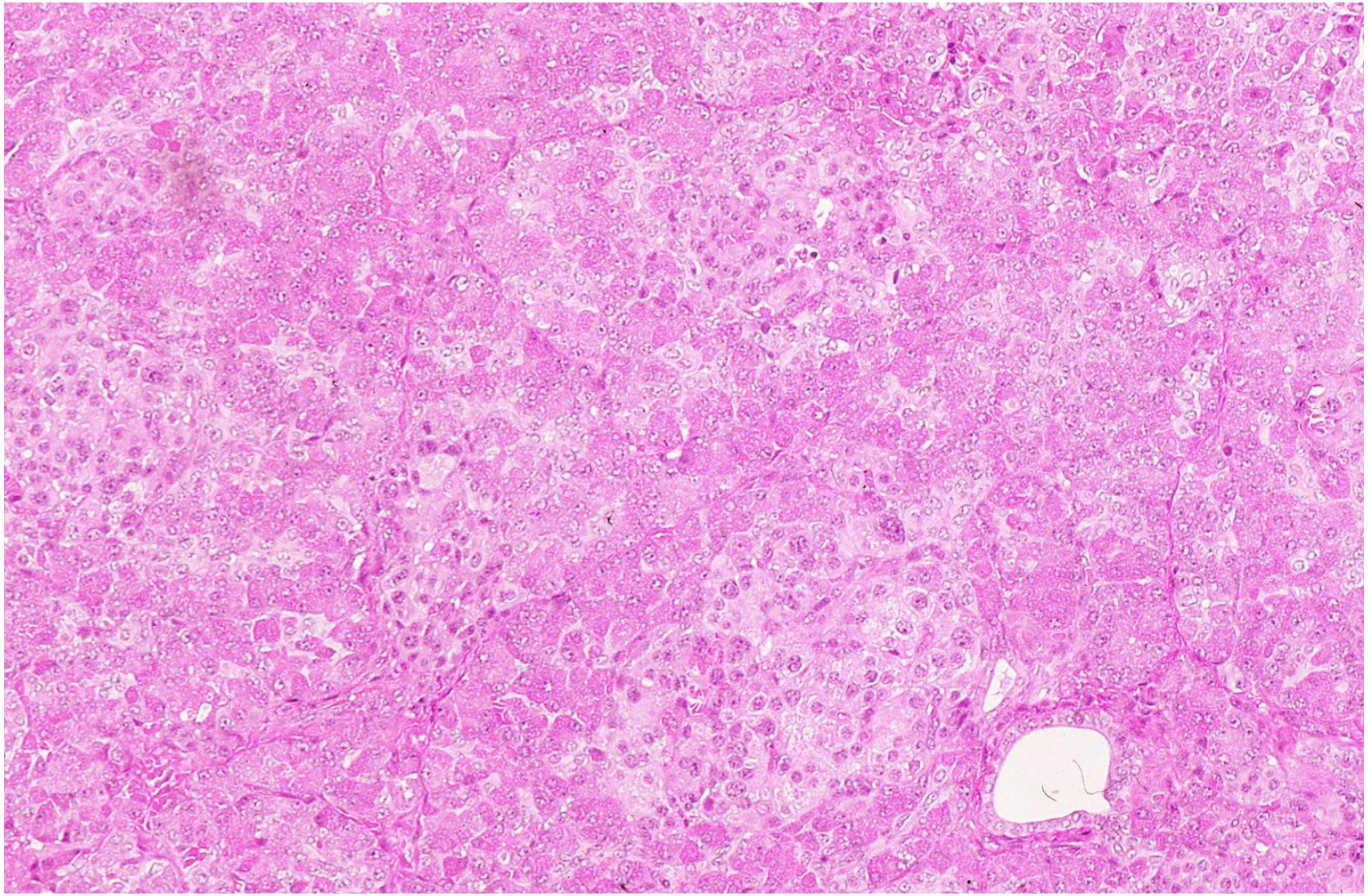
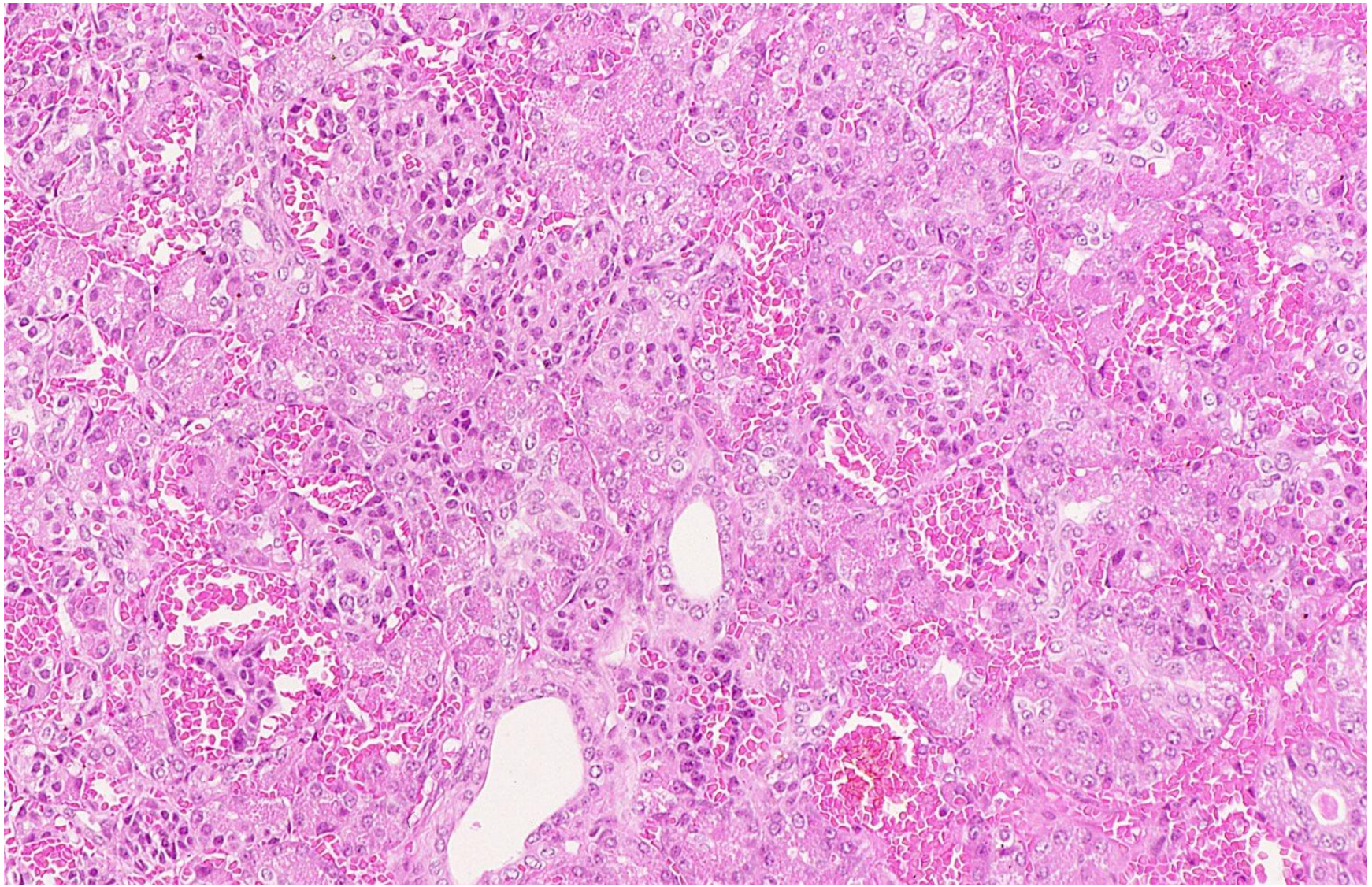
Neonatal nesidioblastosis

Neonatal nesidioblastosis is now called as persistent hyperinsulinemic hypoglycemia of infancy. Most cases occur sporadically, while several genetic defects (SUR1, Kir6.2, GCK, and GLUD1 genes) have been described in the neonates. Biochemical tests for the diagnosis include 1) a supervised fasting test by determining the glucose requirements to maintain normoglycemia, 2) inappropriately high insulin and C-peptide levels, 3) low free fatty acid and ketone body concentrations, 4) glycemic response to glucagon, and 5) the absence of ketonuria. Histopathological features include hypertrophic islets containing beta-cells with pleomorphic nuclei, ductuloinsular complexes and neoformation of islets from ducts. 95% pancreatectomy is necessary in neonates with a diffuse form of nesidioblastosis, whereas focal forms can be treated by partial pancreatectomy.

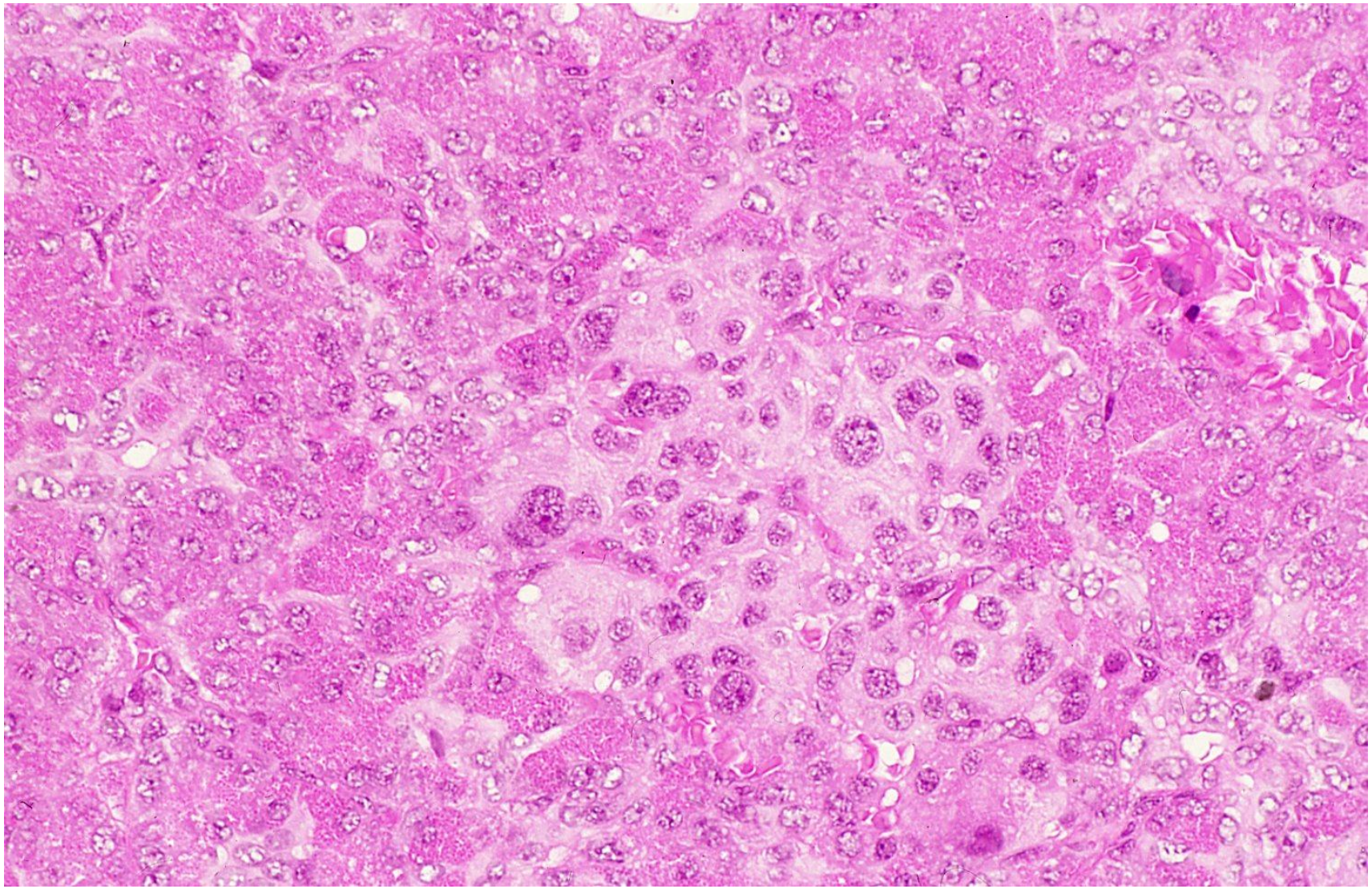
Ref.: Kaczirek K, Niederle B. Nesidioblastosis: an old term and a new understanding. World J Surg 2004; 28(12): 1227-1230. doi: 10.1007/s00268-004-7598-7



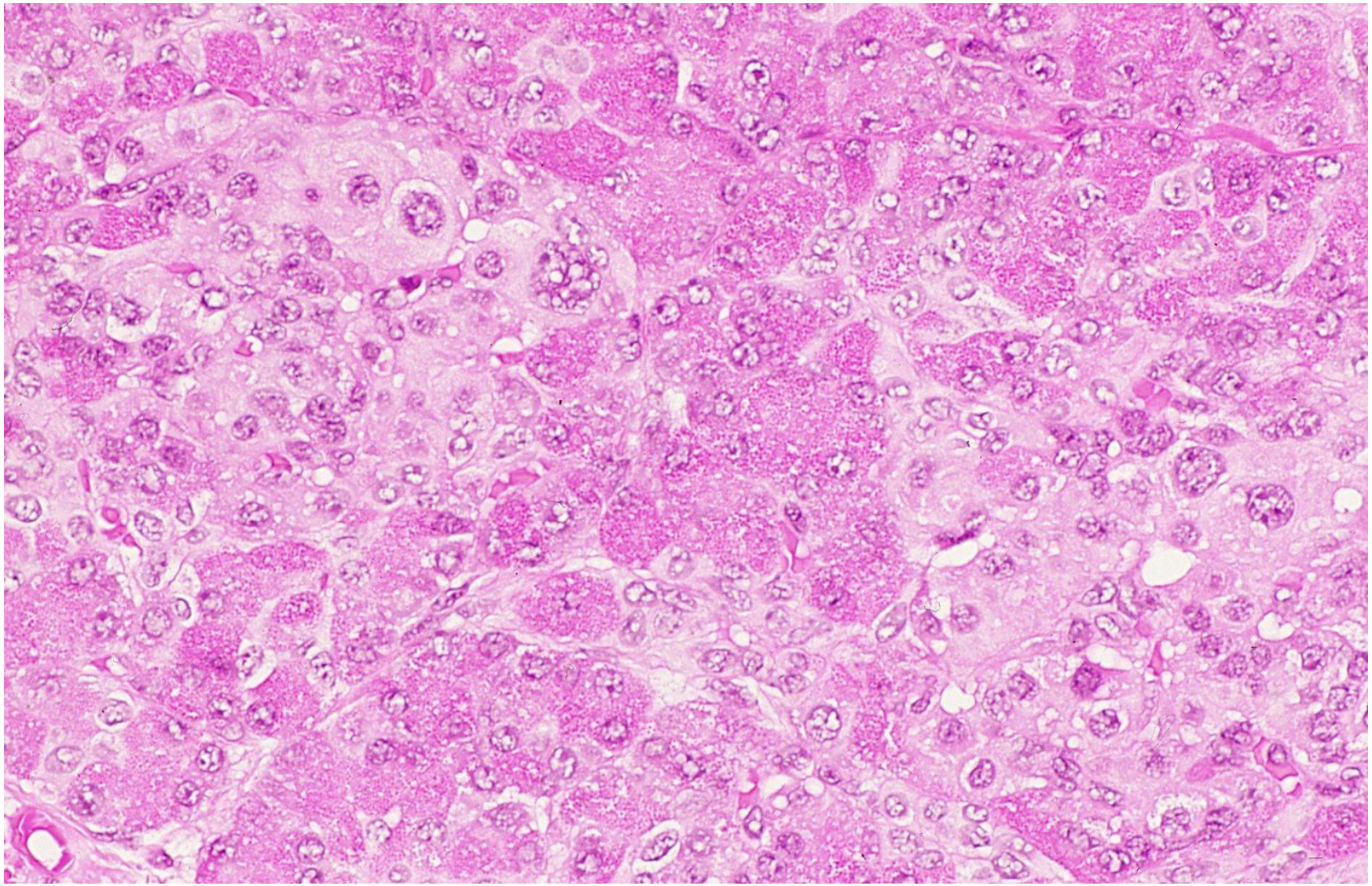
Neonatal diffuse nesidioblastosis (persistent hyperinsulinemic hypoglycemia of infancy) seen in a 2 months-old boy (H&E-1). The 95% pancreatectomy specimen show marked hyperplasia of the islets. Nuclear enlargement is seen in the islet cells with finely eosinophilic cells (beta-cells).



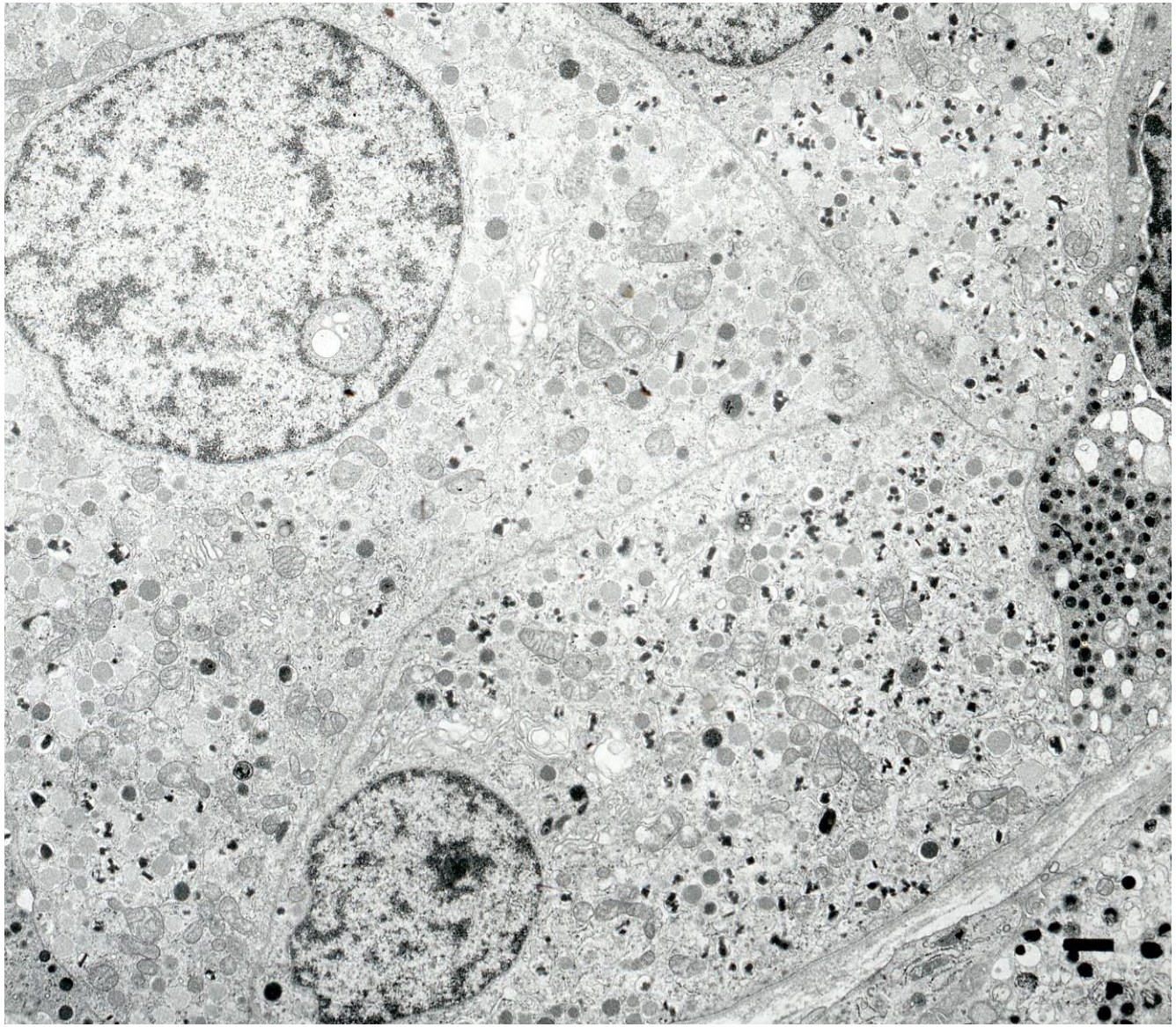
Neonatal diffuse nesidioblastosis (persistent hyperinsulinemic hypoglycemia of infancy) seen in a 2 months-old boy (H&E-2). The 95% pancreatectomy specimen show marked hyperplasia of the islets. Nuclear enlargement is seen in the islet cells with finely eosinophilic cells (beta-cells).



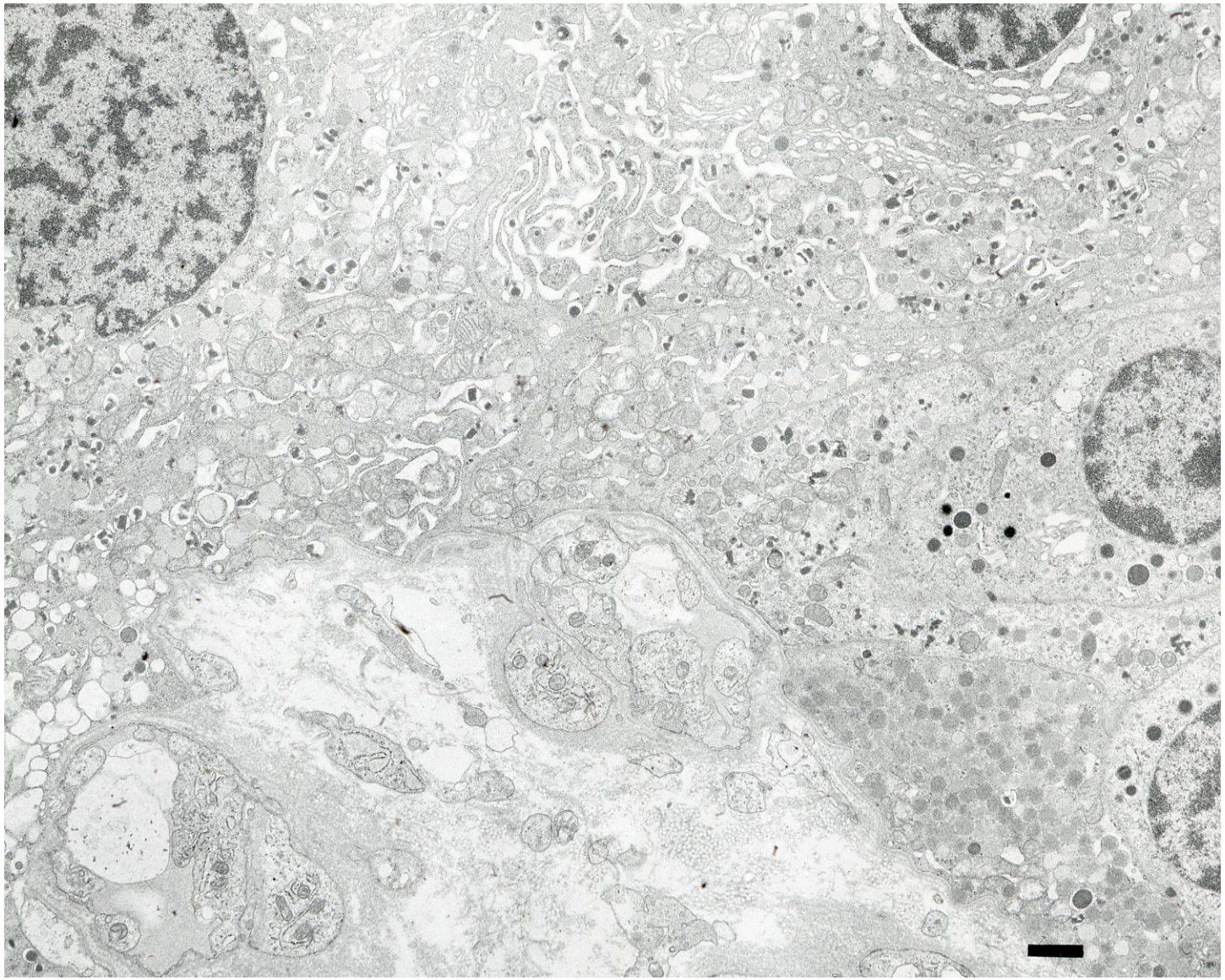
Neonatal diffuse nesidioblastosis (persistent hyperinsulinemic hypoglycemia of infancy) seen in a 2 months-old boy (H&E-3). The 95% pancreatectomy specimen show marked hyperplasia of the islets. Nuclear enlargement with bizarre nucleation is seen in the islet cells with finely eosinophilic cells (beta-cells). Intranuclear cytoplasmic inclusions are focally noted in the islets.



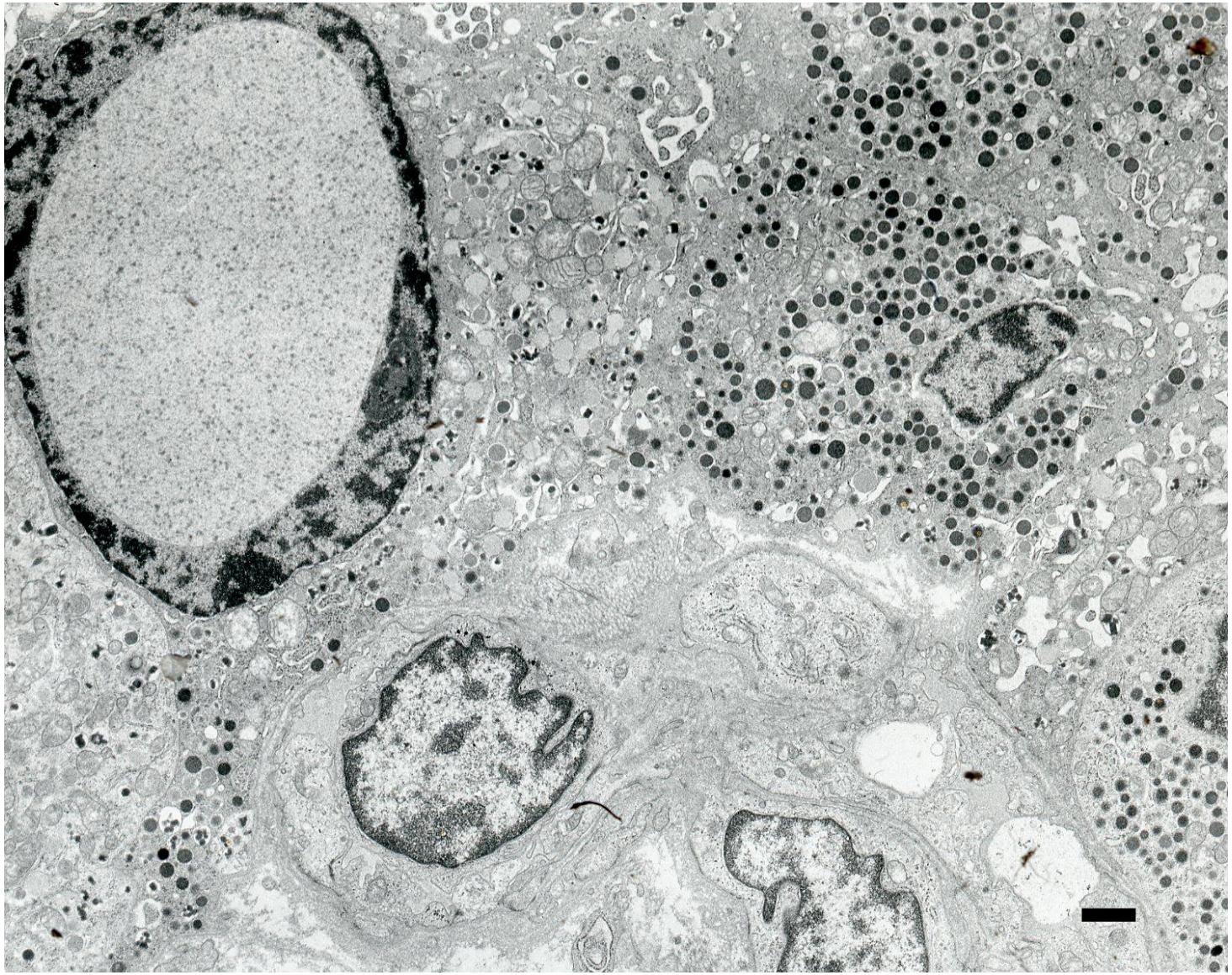
Neonatal diffuse nesidioblastosis (persistent hyperinsulinemic hypoglycemia of infancy) seen in a 2 months-old boy (H&E-4). The 95% pancreatectomy specimen show marked hyperplasia of the islets. Nuclear enlargement with bizarre nucleation is seen in the islet cells with finely eosinophilic cells (beta-cells). Intranuclear cytoplasmic inclusions are focally noted in the islets.



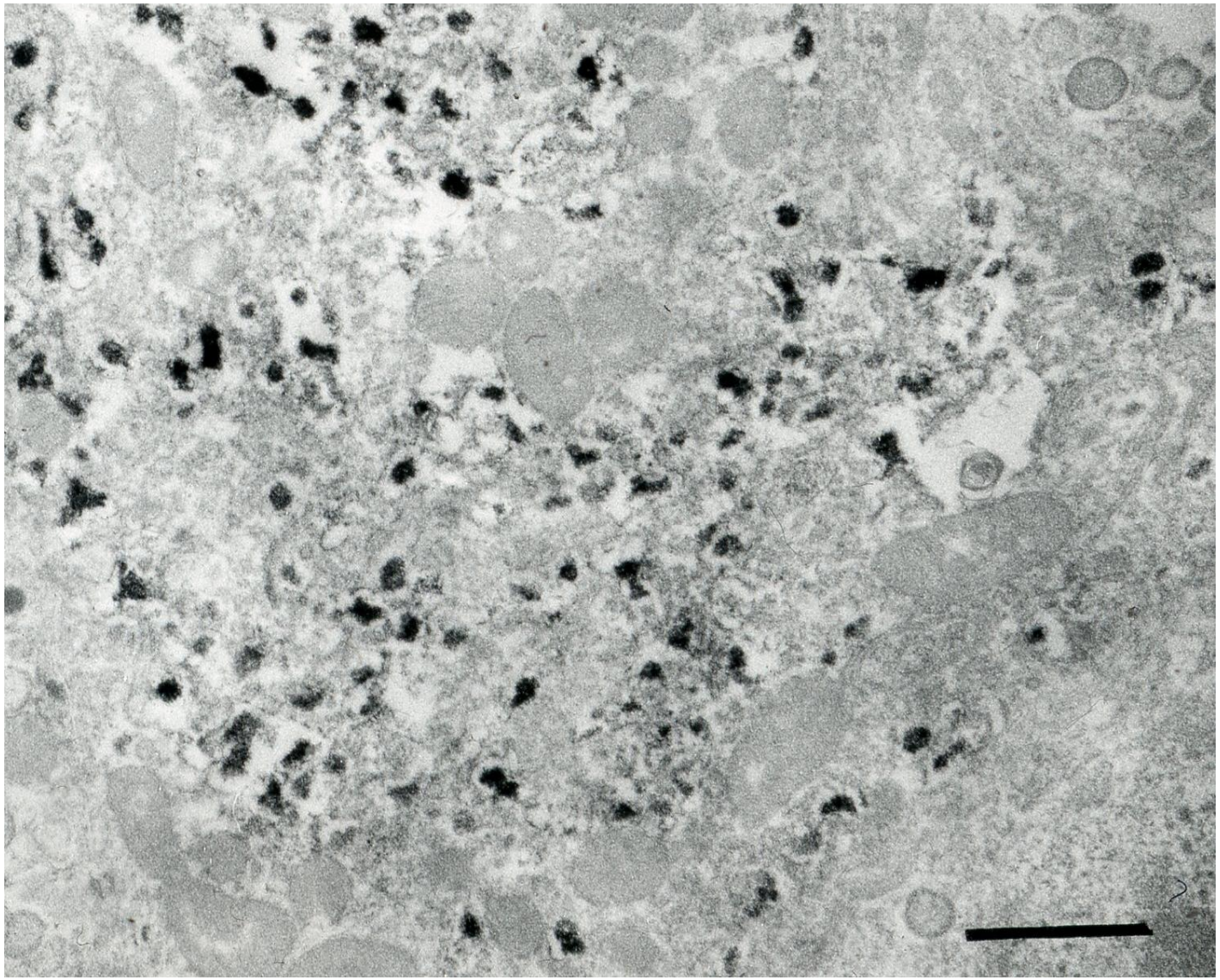
Ultrastructure of neonatal diffuse nesidioblastosis (persistent hyperinsulinemic hypoglycemia of infancy) seen in a 2 months-old boy (EM-1). Islet cells containing beta granules are hyperplastic. A small intranuclear cytoplasmic inclusion is observed in the beta cells.



Ultrastructure of neonatal diffuse nesidioblastosis (persistent hyperinsulinemic hypoglycemia of infancy) seen in a 2 months-old boy (EM-2). Islet cells containing beta granules are hyperplastic. Rough endoplasmic reticula are well developed. Electron-dense round granules in glycagon cells seem to be decreased. Capillary vessels are located just adjacent to the islet cells.



Ultrastructure of neonatal diffuse nesidioblastosis (persistent hyperinsulinemic hypoglycemia of infancy) seen in a 2 months-old boy (EM-3). Islet cells containing beta granules are hyperplastic. A large intranuclear cytoplasmic inclusion with little organelles is observed in the beta cells.



Ultrastructure of neonatal diffuse nesidioblastosis (persistent hyperinsulinemic hypoglycemia of infancy) seen in a 2 months-old boy (EM-4). Higher-powered view of the islet cell containing beta granules is shown. Crystalloid granule cores are evident.