

**Final anatomical diagnosis :**

1. Neonatal hematochromatosis with iron deposits in liver, pancreas, testis, thyroid, small intestine, lung, thymus, spleen, kidney, and adrenal glands and with cardiomegaly.
2. Immaturity of bilateral lungs with no expansion of alveolar spaces.
3. Ascites, serous, 10 C.C.
4. Pleural effusion, serous, right, 10 C.C., left 5 C.C.
5. Pericardial effusion, serous, 3 C.C.

**Clinical summary :**

This 3 D/O male newborn with a history of intrauterine growth retardation was suspected to have patent ductus arteriosus and atrial septal defect. Imaging studies, including brain ultrasound and X-ray, revealed cardiomegaly and intraventricular hemorrhage. His condition was poor with respiratory distress and seizures. He died on January 27, 2000.

**Autopsy findings :**

(1) Body and external appearance :

Weight	2250 GM
Length	49 CM
Development	Poor
Nutritional state	Poor

For infant

Circumference			
Head	33 CM	Thorax	30 CM
Abdomen	30 Cm	Crown to rump	33 CM

(2) Body cavities :

Pleural	Serous effusion	Right : 10 C.C.
		Left : 5 C.C.
Peritoneal	Serous effusion, 10 C.C.	
Pericardial	3 C.C.	

(3) Cardiovascular system :

Greater vascular structures	Gross	Microscopic Diagnosis
	Unremarkable	Unremarkable

Heart General :			
Weight	25.0 GM	Pulmonary valve	2.5 CM
Left ventricle	0.5 CM	Mitral valve	3.0 CM
Right ventricle	0.5 CM	Aortic valve	2.5 CM
Tricuspid valve	3.0 CM	Foramen ovale	Patent
Ductus arteriosus	Patent		

Cardiovascular system	Gross	Microscopic Diagnosis
Valves	Unremarkable	Unremarkable
Epicardium	Unremarkable	Unremarkable
Myocardium	Unremarkable	Few iron deposits in myocytes
Endocardium	Unremarkable	Unremarkable
Coronary arteries	Unremarkable	Unremarkable

(4) Respiratory system :

Trachea and major bronchi	
Gross	Contents : Unremarkable
	Mucosa : Unremarkable
Microscopic Diagnosis	Unremarkable

Lung						
Gross	Weight	Right	15.0 GM	Size	Right	4.5 x 2.5 x 1.8 CM
		Left	9.3 GM		Left	4.4 x 4.2 x 2.1 CM
Microscopic Diagnosis	1. No expansion of alveolar spaces and severe congestion . 2. A few iron deposits.					

Mediastinum / Thymus		
Gross	Weight	2.9 GM
	Size	4.0 x 1.5 x 0.7 CM
Microscopic Diagnosis	A few iron deposits in Hassall's corpuscles.	

(5) Gastrointestinal Tract :

Gastrointestinal Tract	Gross	Microscopic Diagnosis
Pharynx	Unremarkable	Unremarkable
Esophagus	Unremarkable	Unremarkable
Stomach	Unremarkable	Unremarkable
Duodenum	Unremarkable	Moderate iron deposits
Small bowel	Unremarkable	Moderate iron deposits
Large bowel	Marked dilatation of rectum and sigmoid colon	Unremarkable
Cecum and appendix	Unremarkable	Unremarkable

Pancreas		
Gross	Weight	1.8 GM
	Size	3.5 x 0.8 x 0.6 CM
Microscopic Diagnosis	Heavy iron deposits in Acini and islands of Islet.	

(6) Hepatobiliary system :

Liver		
Gross	Weight	44.4 GM
	Size	6.0 x 4.0 x 3.5 CM
	Out surface	Multifocal collapsed
	Cut surface	Green and soft with multifocal hemorrhage, necrosis and cystic change.
Microscopic Diagnosis	1. Severe iron deposits in liver cells, bile ducts and Kupffer cells. 2. Severe cholestasis. 3. Multifocal hemorrhage or coagulative necrosis.	

Gallbladder, Bile ducts	
Gross	Unremarkable
Microscopic Diagnosis	Unremarkable

(7) Hematopoietic-Lymphoid system :

Spleen		
Gross	Weight	10.5 GM
	Size	4.5 x 2.5 x 1.85 CM
Microscopic Diagnosis	Marked congestion	

Lymph nodes	
Gross	Unremarkable
Microscopic Diagnosis	Unremarkable

(8) Urogenital system :

Kidney						
Gross	Weight	Right	8.2 GM	Size	Right	3.2 x 2.5 x 1.8 CM
		Left	9.7 GM		Left	4.0 x 2.2 x 2.0 CM
	Unremarkable					
Microscopic Diagnosis	Few iron deposits in proximal tubules and without tubular dysgenesis.					

Urogenital system	Gross	Microscopic Diagnosis
Ureters	Unremarkable	Unremarkable
Bladder and urethra	Unremarkable	Unremarkable
Testes	Unremarkable	Iron deposits in Sertoli and Leydig cells

(9) Endocrine system :

Thyroid		
Gross	Weight	0.34 GM
	Unremarkable	
Microscopic Diagnosis	Heavy iron deposits in follicular cells.	

(10) Urogenital system :

Adrenals						
Gross	Weight	Right	1.7 GM	Size	Right	2.5 x 2.0 x 1.0 CM
		Left	1.9 GM		Left	3.0 x 1.4 x 1.0 CM
Microscopic Diagnosis	Few iron deposits in cortex					

**Section Taken and Labeled :**

Labeled	Section Taken	Labeled	Section Taken
A	Bone marrow	K	Spleen
B	Left testis	L	Pancreas (缺蠟塊)
C	Right testis	M	Thymus
D	Left adrenal gland	N	Thyroid gland
E	Right adrenal gland	O	Trachea and larynx
F	Left kidney	P	Stomach and small intestine
G	Right kidney	Q	Sigmoid colon and rectum
H	Urinary bladder	R	Right ventricle of heart
I	Left lung	S	Left ventricle of heart
J	Right lung	T1-6	Liver (T2, T4 缺蠟塊)

**Final Comments :**

The newborn showed extensive iron deposits in liver, pancreas, thyroid, testis, small intestine, heart, kidney and adrenal glands. It could be a case of neonatal hematochromatosis. There are also multifocal hemorrhagic necrosis of liver and severe cholestasis. It is not associated with renal tubular dysgenesis.