

## CASE 1

### CLINICAL HISTORY

This 53 year-old woman first presented with aortic valve disease due to aortic root involvement by Still's Disease (juvenile rheumatoid arthritis), first diagnosed in 1971. On long term immunosuppression since then. She underwent homograft aortic valve replacement in 1976, which required replacement because of degeneration, together with replacement of the aortic root in 1992, with a third re-do valve replacement in 2006. A patent foramen ovale was repaired during her first aortic valve replacement. Also known to have heart rhythm problems - left bundle branch block, episodes of ventricular ectopics - from her post-operative course during one of her previous cardiac surgical episodes.

She developed a dental abscess in March 2006 which was dealt with by extraction of the tooth and treatment with antibiotics but subsequently developed worsening aortic regurgitation with a dilating left ventricle, presumed due to infective endocarditis although it is not known if any organism was grown.

On 04/08/08 she underwent homograft aortic valve replacement using a Mitroflow tissue valve. Initially had a raised creatinine but this settled without haemofiltration. Also had bi-basal collapse and consolidation of both lungs on chest x-ray, managed appropriately. Required beta blockers for tachycardia. On day 8 post-operatively she suffered a PEA cardiac arrest and died.

Past medical history also includes innumerable operations on all her joints, emergency laparotomies for perforated duodenal ulcer, emergency appendectomy. She also had a pharyngeal pouch with dysphagia, making it difficult to maintain her body weight (BMI 16). Known to have very poor mouth opening limited to 1cm, severely restricted neck movement and an unstable odontoid peg, all increasing her anaesthetic risk.

## **SUMMARY OF AUTOPSY FINDINGS**

Height: 1.6m      Weight: 41.5kg      BMI: 16

### **RESPIRATORY SYSTEM**

There was 200ml of blood stained effusion in the right thoracic cavity. The larynx, trachea and major bronchi contained a small amount of froth and mucous. Both lungs were moderately oedematous. There was blood clot surrounding a small tear in visceral pleura overlying posterior segment of right upper lobe. No pulmonary thromboemboli were found.

### **CARDIOVASCULAR SYSTEM**

The pericardium was open anteriorly but adherent to epicardium by fibrinous adhesions. There was no significant effusion.

The heart weighed 476g. There was mild biventricular hypertrophy but no focal lesions were seen. There was biventricular dilatation. Dense scarring was noted in endocardium on both sides of atrial septum.

There was calcification of the mitral valve at lateral commissure, and extending onto anterior cusp and left site of atrial septum adjacent to the medial commissure. There was a patch of calcification on the anterior cusp of TV. The tissue AV (stented) was satisfactorily in place. The pulmonary valve was normal. The coronary arteries were right dominant and were normal as were the coronary ostia.

The aortotomy and incision sites were secure. The aortic root and ascending aorta was heavily scarred with intimal fibrosis and calcification relating to previous aortotomy and incision sites. Pulmonary artery was normal and no pulmonary thromboembolus was found. The vena cava and other veins were normal.

## **GASTROINTESTINAL SYSTEM**

There was a 25mm diameter pharyngeal pouch originating from the posterior wall of pharynx at its junction with oesophagus and slightly to the left of midline. The opening into the oesophagus itself was narrow but could be easily probed. The pouch was empty. The appendix was absent. The liver was normal, weight 1264g. The remainder of the gastrointestinal system was normal.

## **GENITOURINARY SYSTEM**

Both kidneys showed granular external surfaces, the left including some pitted scars. There were several small yellow nodules, maximum 3mm diameter, scattered over the surface of both kidneys. The remainder of genitourinary system was normal.

## **ENDOCRINE SYSTEM**

Both lobes of thyroid appeared shrunken and fibrotic with a small nodule at the upper pole of right lobe.

## **LYMPHORETICULAR SYSTEM**

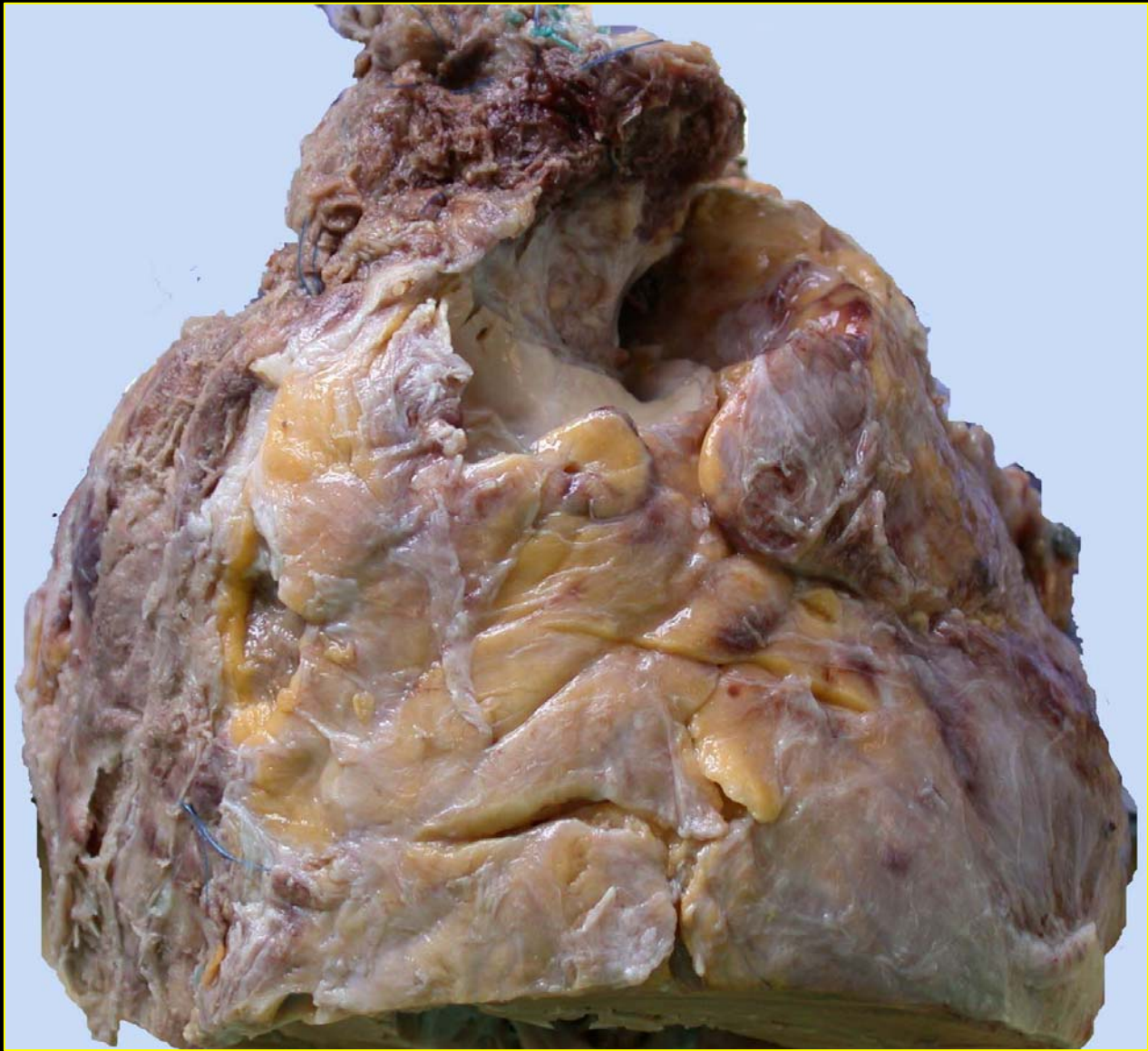
The spleen weighed 186g and was normal. There was generalised lymphadenopathy, discrete rubbery nodes being noted in mediastinum, epigastrium, mesentery and retroperitoneum.

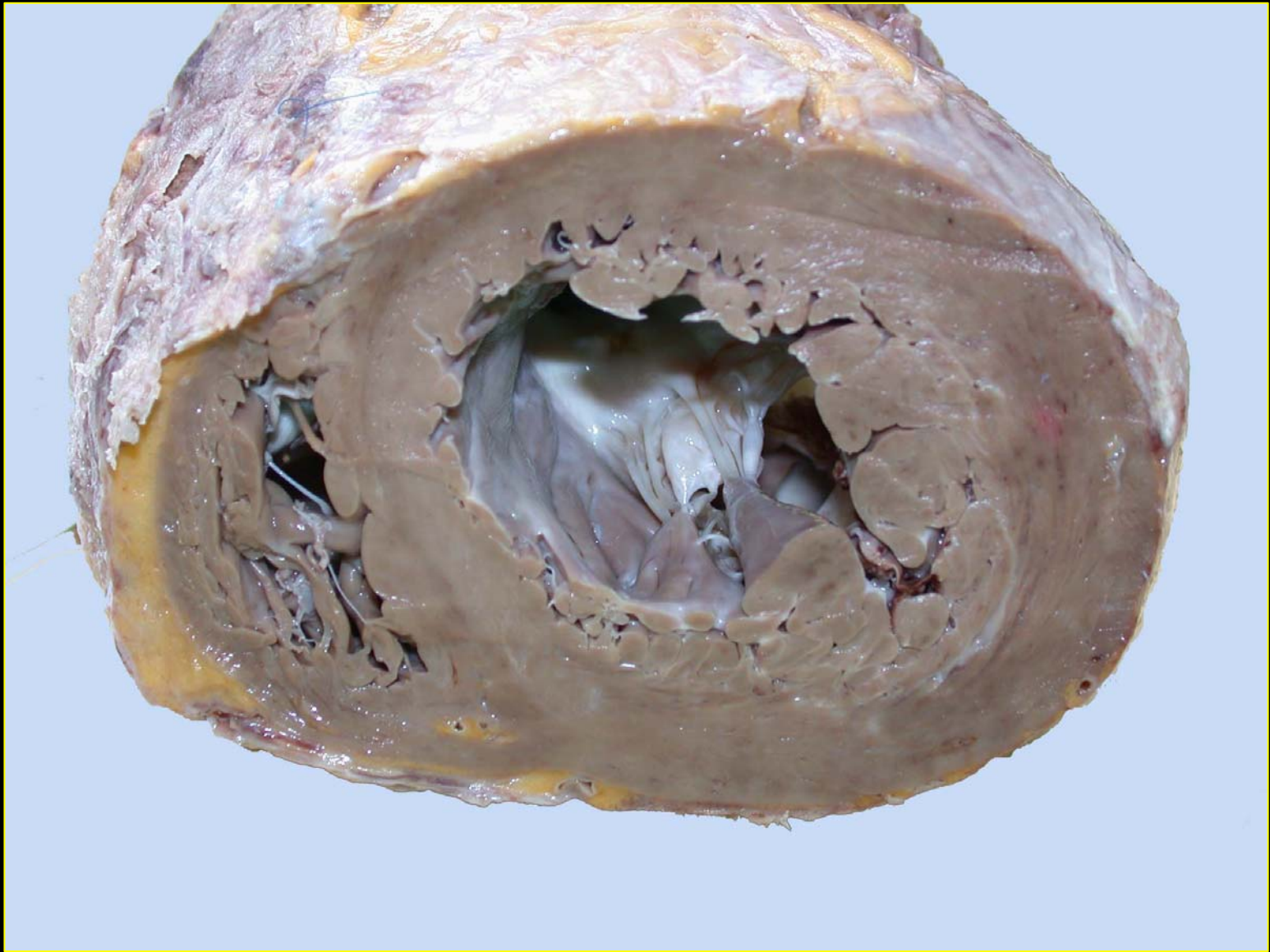
## **CENTRAL NERVOUS SYSTEM**

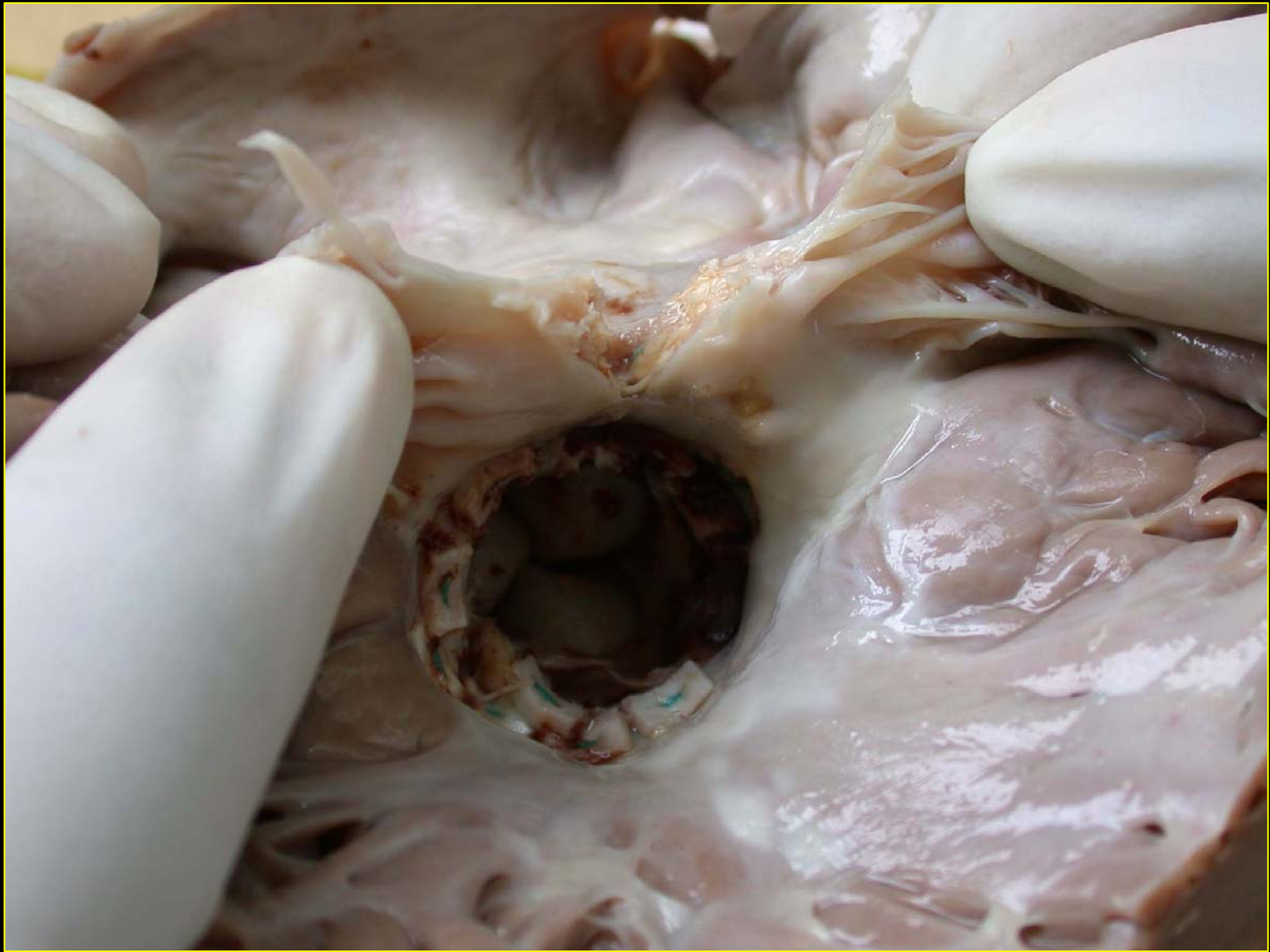
No significant abnormality.

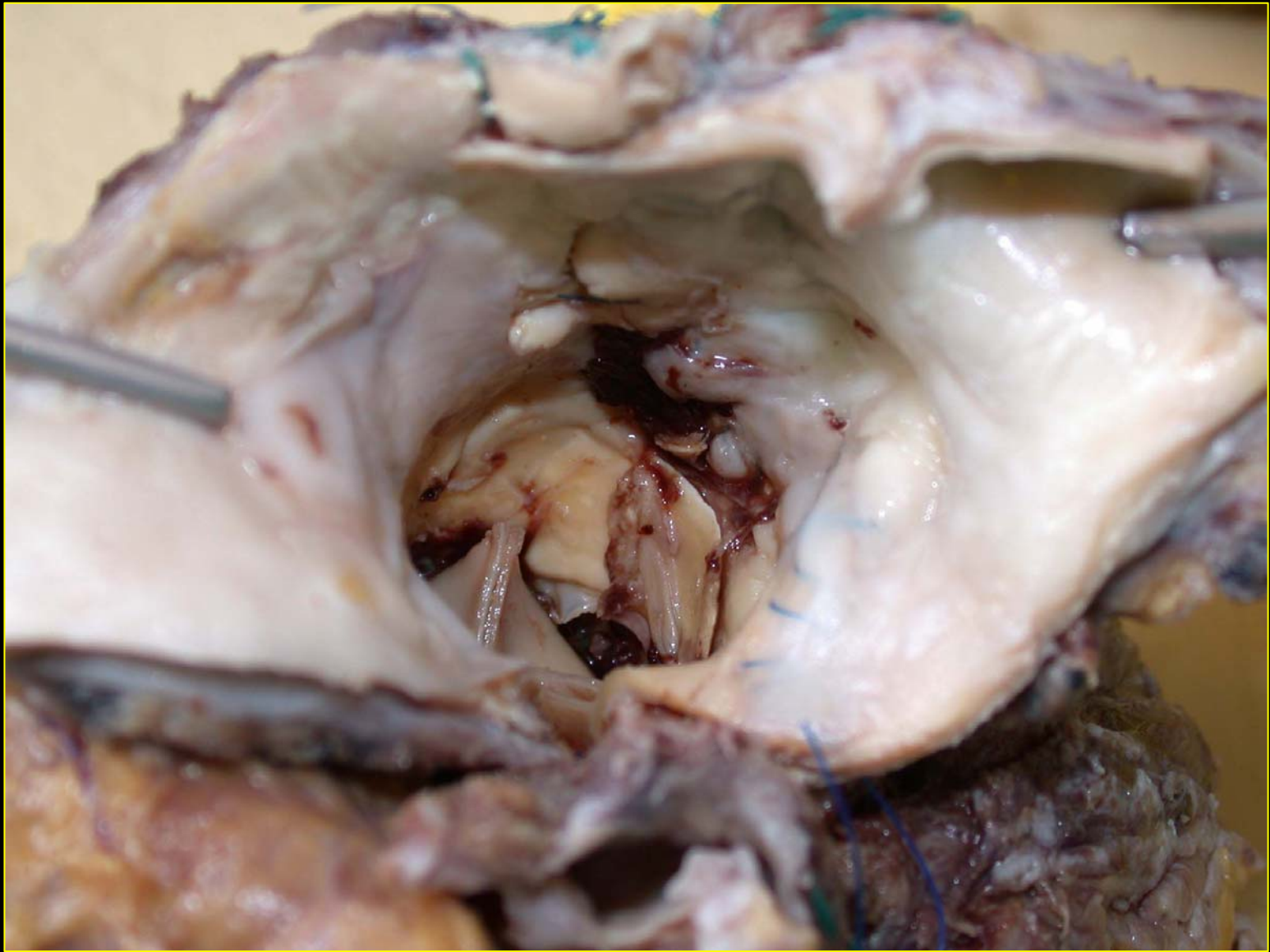
## **SECTIONS SUBMITTED**

Heart [lateral left ventricle, septum] with Elastic Van Gieson stains on both blocks, kidney, representative lymph node

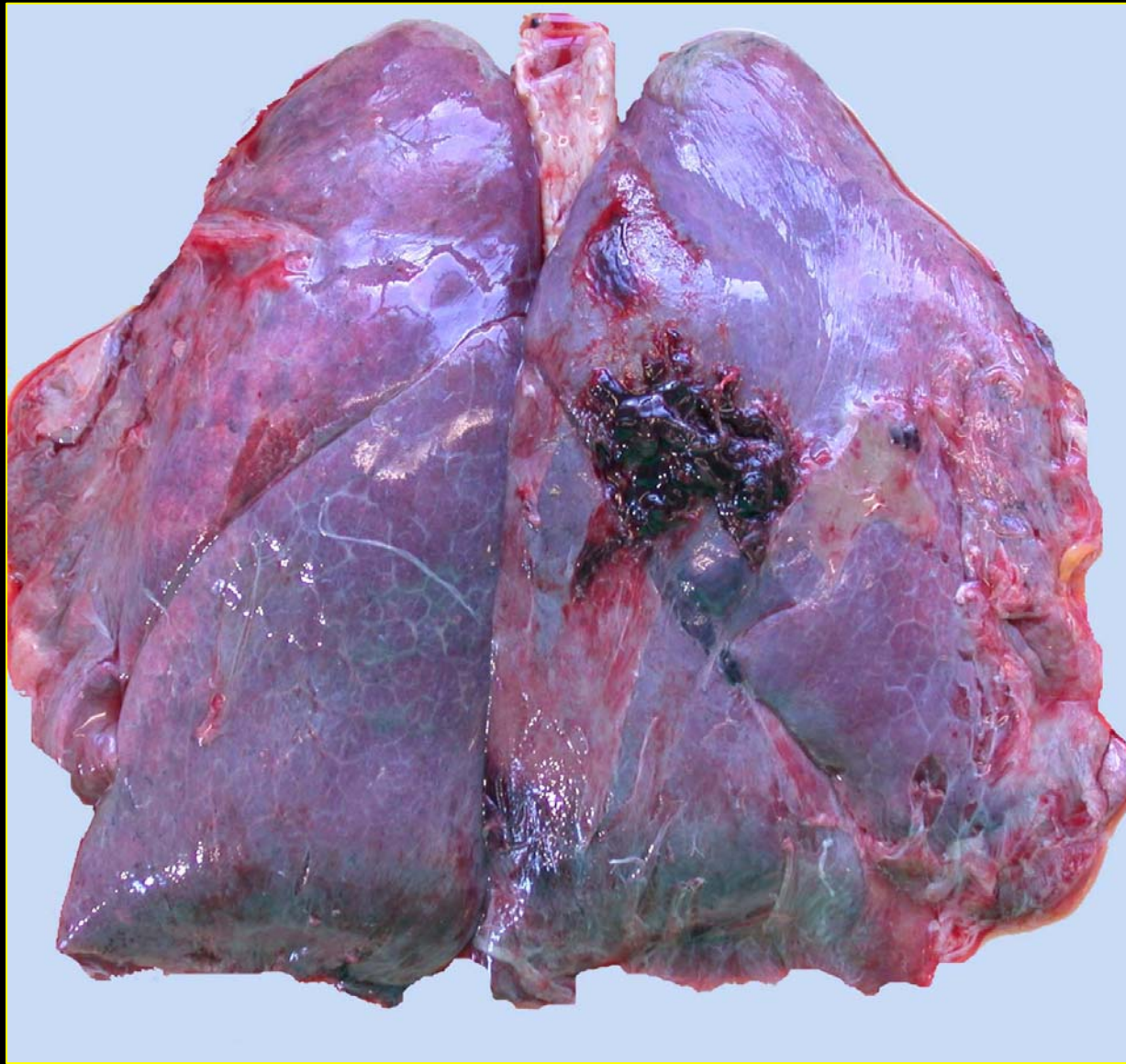


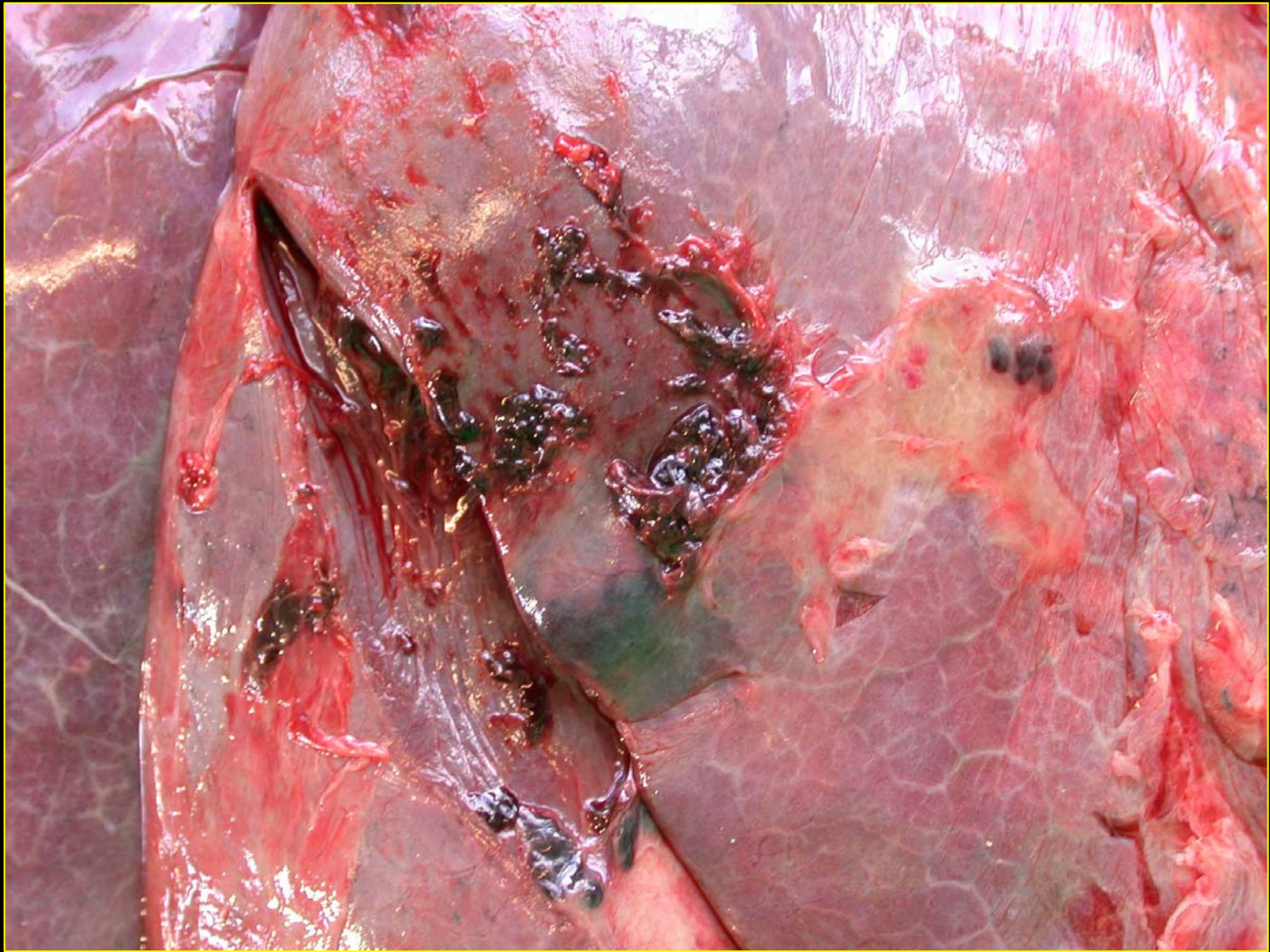












*Pharyngeal pouch*

