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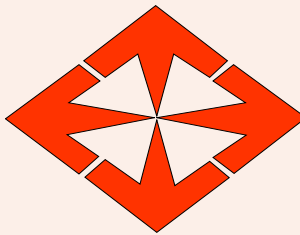
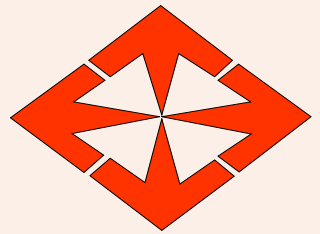
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Outcomes of Liver Transplantations in Patients with Alagille Syndrome with high Pulmonary Arterial Pressure Performed in Our Center



Adem ŞAFAK¹, Emre KARAKAYA¹, Birgül VARAN², Figen ÖZÇAY³, Nedim ÇEKMEN⁴, Sedat YILDIRIM¹, Mehmet HABERAL¹

¹Baskent University, Department of General Surgery, Division of Transplantation

²Baskent University, Department of Pediatric Cardiology

³Baskent University, Department of Pediatric Gastroenterology

⁴Baskent University, Department of Anesthesiology

INTRODUCTION

Alagille syndrome (AGS) is an **autosomal recessive** disorder with multisystemic involvement. Patients with Alagille syndrome are accompanied by many anomalies such as atypical facial appearance, posterior embryotoxon, butterfly vertebra, **cardiac anomalies**, and **biliary problems**.

Cardiac anomalies make intraoperative and postoperative patient management **difficult in patients requiring liver transplantation**. In this study, we present 10 cases of liver transplantation due to Alagille syndrome.

Biliary Problems

Butterfly
Vertebrae

Posterior
Embryotoxon

Alagille syndrome (AGS)

Atypical Facial Appearance

Prominent forehead

Deep set eyes

Bulbous tip of nose

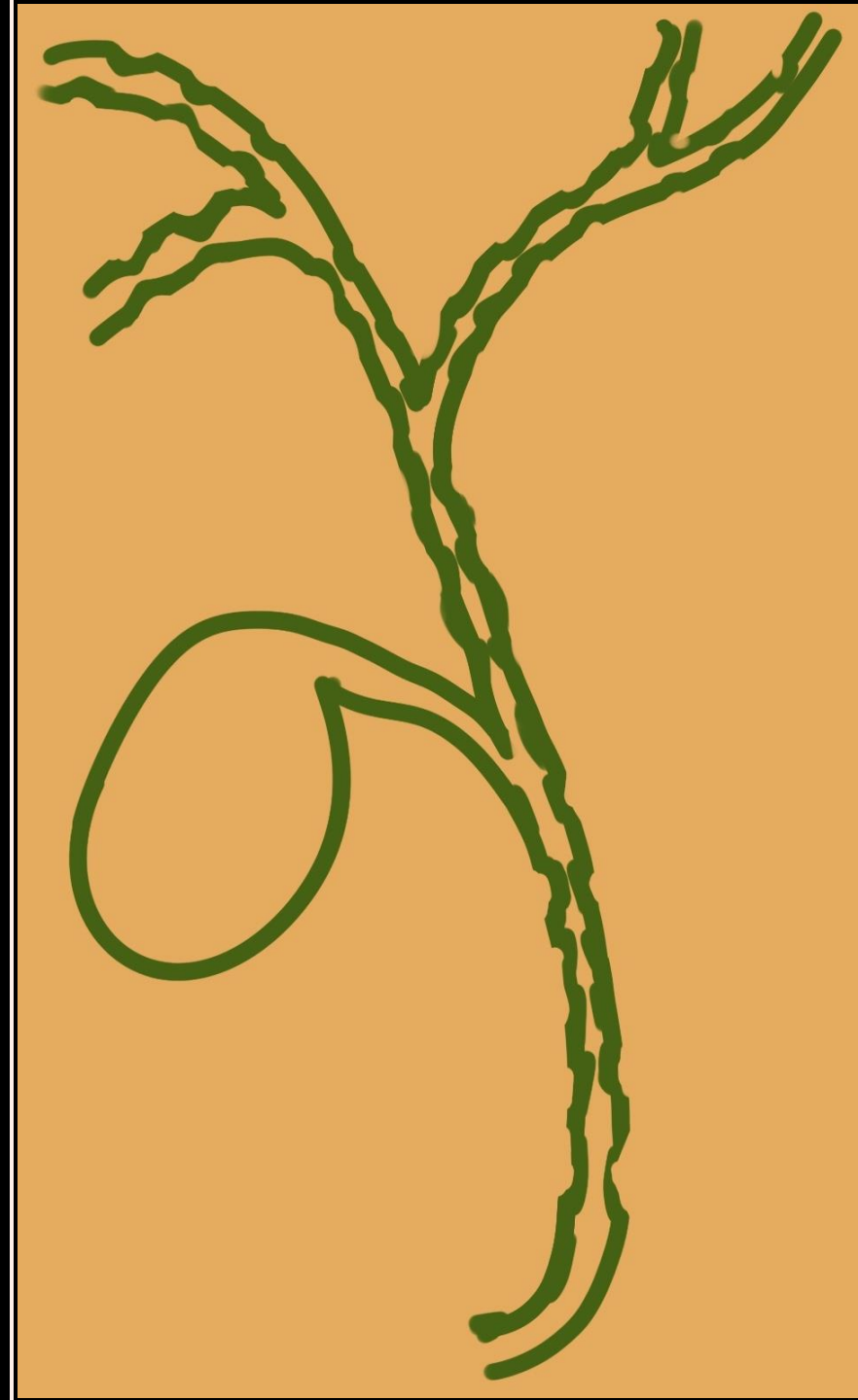
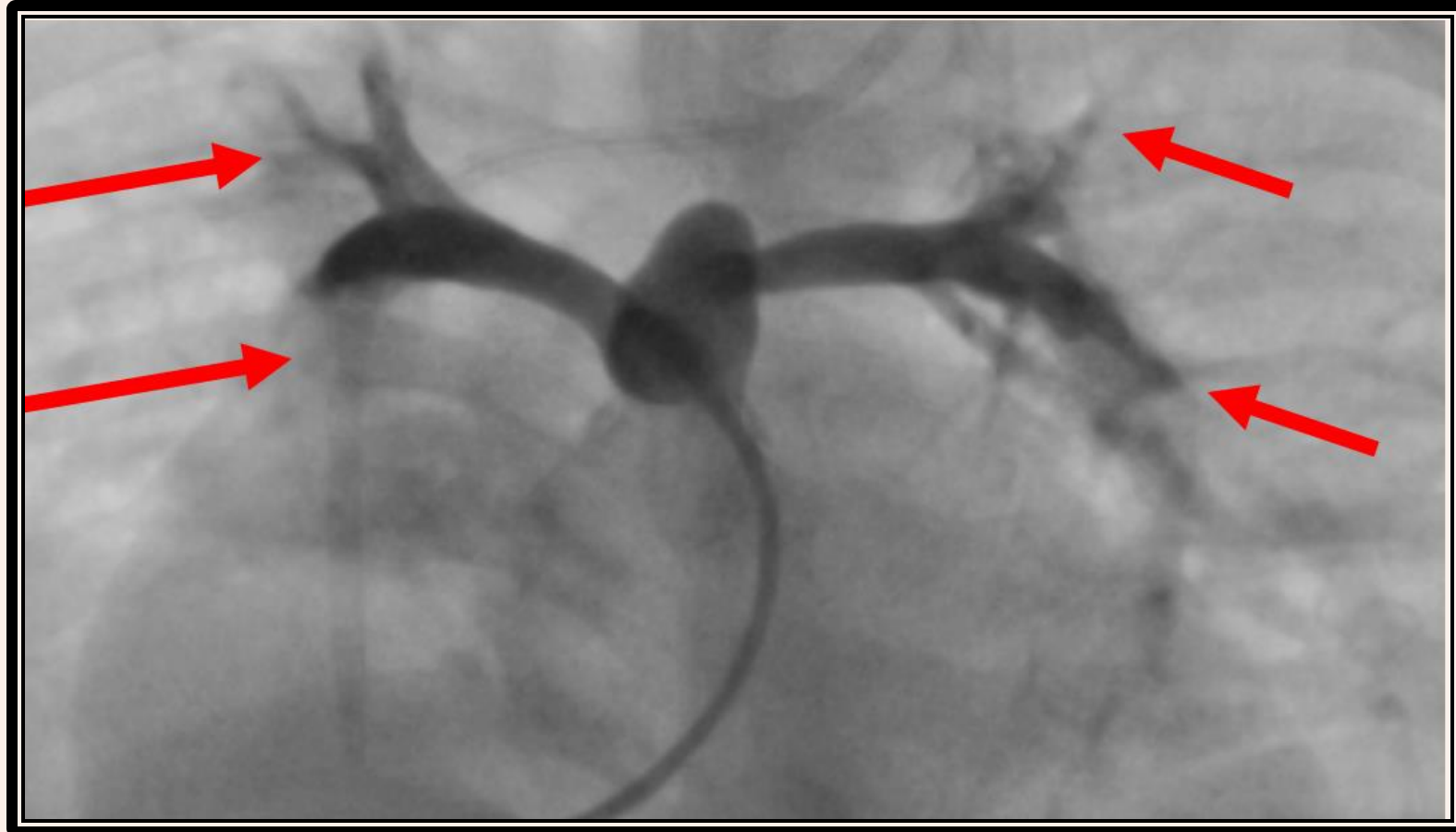
Pointed chin



Renal
Disease

**Cardiopulmonary
Anomalies**

The most important involvement that determines mortality is **bile duct deficiency** and **peripheral pulmonary stenosis.**



The most important involvement

PULMONARY ARTERY PRESSURE

<25: normal

25-35:mild

35-45:moderate

45>:severe

PATIENTS

745 liver transplant

366 pediatric, 379 adults

10 pediatric patients had
alagille syndrome

PARAMETERS

Morphological findings,

**Cardiopulmonary
pathology**

Perioperative complications

RESULTS

8/10 Patients: Peripheral Pulmonary Stenosis

The Highest Right Ventricular Pressure: **110mmHg**

2/8 Patients: Preoperative Pulmonary Angioplasty

0 Patient: Perioperative Complications

0 Patient: Early Mortality

CONCLUSION

1. Since it has multisystemic involvements, it should be evaluated multidisciplinary. (*anesthesia, cardiology, nephrology, gastroenterology, surgery...*)
2. All patients should first be evaluated with **echocardiography**.
3. Cardiac anomalies should be determined, and **pulmonary catheterization** should be performed if necessary.
4. Intraoperative volume balance should be closely monitored (**especially in the anhepatic phase**).