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Aortic coarctation and infantile hemangioma: coincidence or PHACES syndrome?

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Objective: To determine the prevalence of PHACES syndrome (Posterior fossa malformations, Hemangiomas, Arterial anomalies, Cardiac defects, Eye anomalies, Supraumbilical raphe and/or Sternal pit) in patients with obstructive aortic arch pathology (OAAP) and to achieve more insight in the possible association between infantile hemangiomas (IH) and cardiovascular anomalies.

Study design: Pediatric patients diagnosed between 1999 and 2013 with OAAP in our tertiary referral center were included. Questionnaires focusing on the (past) presence of an IH and other symptoms fitting the diagnostic criteria of PHACES syndrome were designed. Data of deceased patients were analyzed separately.

Results: Questionnaires send to 286 patients with OAAP, were returned by 175 subjects (response rate 66%). In 9 cases an IH was diagnosed. One child met the criteria of PHACES syndrome. This child demonstrated a segmental hemangioma and an atypical interrupted aortic arch with atresia of the left common carotid artery, which fits the complex vasculopathy seen in PHACES syndrome. All other children did not meet the PHACES criteria due to characteristics of IH or aortic arch and thus were thought to be normal distribution. Deceased children suffered from severe congenital anomalies though none of them fitted the PHACES syndrome criteria.

Conclusion: In this retrospective cohort, one child met the PHACES criteria, indicating that PHACES syndrome in AC patients is less common than expected. Our study was not able to provide further evidence for the suggested association between IH and obstructive aortic arch pathology.