feature which is almost pathognomonic. Walter *et al*[8] also demonstrated similar stellate KPs in CMV uveitis due to fibrin deposition around inflammatory cells. Under specular microscopy in patients with PSS, Pillai *et al*[9] also observed a similar fibrin deposition around an individual KP resulting in a large conglomeration, giving rise to the classical 'stellate' appearance. These similarities further support a common aetiology between PSS and CMV or herpetic keratouveitis.

Conclusion

We postulate that PSS is not a distinct entity but may represent a spectrum of inflammatory responses to members of the herpesviridae family including CMV and HSV. An acute relapse may present with classic hypertensive cyclitis with or without other clinical manifestations of anterior segment inflammation indistinguishable from or even pathognomonic for 'herpetic' infections including corneal endotheliitis. This observation has implications on future treatment of this condition. Further investigations are necessary to confirm these postulations in large patient series.

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Sir,

Reply to ocular pathology in congenital heart disease

We read with interest the paper of Mansour *et al*[1] on ocular pathology in congenital heart disease.

In a recently published study,[2] we have focused our attention on the relationship between heart and ocular defects in Down's syndrome (DS) patients.

Our study based on 65 DS patients (aged between 1 month and 15 years old), followed up with an ophthalmological examination at birth and one each year, showed that in 17 cases (26%), congenital heart disease (CHD) and ocular anomalies (OA) were significantly associated (χ^2 test, $P<0.01$).

We also found a recurrent association between nystagmus (4/6) and congenital cataract (3/3) with atrial septal defects and between myopia and severe CHD (three with atrioventricular canal and two with Fallot tetralogy on six cases), suggesting a possible specific pattern of association.

Moreover, Bromham *et al*[3] observed that in children with DS, heart defects were associated with both myopia and nystagmus and not with other ocular anomalies.

We have searched for possible association between ocular and heart anomalies in the Sicilian Registry of Congenital Malformations database and we found 15 cases of nonsyndromic congenital cataract. In five cases (30%; $P=0.45$), CHD was also reported (four with atrial septal defect and one with ventricular septal defect).

This second set of data confirms the hypothesis of a link between congenital cataract and atrial septal defects, even if this type of CHD is common and a causal relationship is difficult to assess with small sample size.

On the basis of these observations, it is possible to reinforce the hypothesis that susceptibility genes for specific CHD and OA may be contiguous or reciprocally influenced.

The study of large patient sample with specific recurrent associations may contribute in the search of possible candidate gene both for CHD and OA. On the other hand, the association between ocular and heart anomalies suggest that a multidisciplinary approach is needed and all CHD patients, syndromic and isolated, may be evaluated routinely by an ophthalmologist.

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Sir,

Reply to Bianca *et al*

The letter of Bianca *et al*[1] has shed light into the association between nystagmus, cataract, or myopia and congenital heart disease (especially atrial septal defect) in the context of Down's syndrome. Nystagmus was detected in 30% of Down's syndrome.[2] da Cunha *et al*[3] found myopia to be associated with congenital heart disease in Down's syndrome, while Bromham[4] found heart defects in Down's syndrome to be associated with myopia and nystagmus.

The series that we described had a small number of Down's syndrome (17 cases) and were examined at a very young age, and many had poor oxygen saturation at the time of the eye examination. Ocular findings increase with age, and because of the cross-sectional nature of the study, we found a low percentage of ocular findings: two had congenital cataract and one had congenital nystagmus among the 17 subjects with Down's syndrome. The high percentage of congenital cataract in Down's syndrome (12%) confirms to the findings of Bianca *et al*.[1]

We analysed the cases of isolated atrial septal defect and found negative eye examination in 14 subjects (including three with velocardiofacial syndrome), ptosis (one subject), and congenital cataract (one subject). It is possible that congenital cataract is associated with atrial septal defect as suggested by Bianca *et al*.[1]

We thank Bianca *et al* for their theory of susceptibility genes for atrial septal defect or other cardiac anomalies and cataract or other eye anomalies that may be contiguous or reciprocally influenced. Larger epidemiological studies than ours can help elucidate these associations.

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