VSD is the second most common defect in trisomy-21 after complete atrioventricular septal defect. Irving and Chaudhari (2) showed in their study, on the basis of a population registry, that among 821 infants born with Down syndrome, 31% had VSD. The short life expectancy in patients with Down syndrome can be related to other noncardiac causes. Uppal et al. (3) showed that respiratory-related comorbidities and dementia are among the leading causes of mortality in Down syndrome. In our study of interest (1), survival rates were compared with the expected survival rates of an age-matched Dutch population. Information on the prevalence of trisomy-21 in both groups was not provided. Although the cumulative survival rate was only slightly lower than the general Dutch population after excluding early post-operative mortality, comparing the study cohort with a matching group with similar prevalence of trisomy-21 will result in more accurate cumulative survival rates.

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REPLY: The Unnatural History of Ventricular Septal Defect

We thank Dr. Marmoush and colleagues for their response to our paper (1) describing the survival and clinical outcome in patients up to 40 years after surgical closure of a ventricular septal defect (VSD).

Dr. Marmoush and colleagues justly remarked the high incidence of VSD in infants with Down syndrome. Patients with Down syndrome have a shorter life expectancy than the general population (2). In our study, we used survival rates of an age-matched Dutch population, which were on the basis of all Dutch residents, including patients with Down syndrome. In the 1980s, the estimated incidence of Down syndrome among newborns in the Netherlands was 10 to 14 per 10,000 live births (3). In our study cohort consisting of 174 patients with VSD, none had Down syndrome, even though Down syndrome was not an exclusion criterion. This could probably be explained by the fact that in this early surgical era heart defects were not always surgically repaired in patients with Down syndrome and by the relatively small number of included patients. Therefore, we think that, regarding the prevalence of Down syndrome, it was justified to compare the survival rates of our study cohort with those of the normal Dutch population. However, 8 patients with other syndromic, chromosomal, or psychomotor abnormalities participated in our study: psychomotor retardation (n = 3), 22q11 deletion (n = 2 of whom 1 deceased), VACTERL (Vertebral anomalies, Anal atresia, Cardiac defects, Tracheoesophageal fistula and/or Esophageal atresia, Renal & Radial anomalies and Limb defects) association (n = 1), Holt-Oram syndrome (n = 1), and hereditary motor and sensory neuropathy (n = 1). The mortality in this subgroup of patients with a syndrome is comparable with the mortality in our complete cohort (i.e., 31 deceased patients out of the 156 traceable patients). In conclusion, we agree that syndromic or chromosomal abnormalities can affect survival rates in general, but this does not affect the results in our study.

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