APPENDIX. CONFERENCE DISCUSSION

The Ross/Konno procedure is described in the current issue of EJCTS, aszyk et al. present their excellent experience with the Konno-Ross operation in 16 infants, primarily having complex left ventricular outflow tract obstruction (LVOTO). I want to specifically focus on this part of the population because you describe in your paper that only 40% of the patients were finally without any medication, so 60% were, as I read it here, on heart failure medication, on diuretics, beta blockers and ACE inhibitors. So I wonder, did you study the pulmonary hypertension on latest echo, or the diastolic behaviour of the left ventricle, because even if these patients are now after five years in validity class 1, one might imagine that some of them have the problem of a stiff, noncompliant left ventricle. And it is, of course, very difficult to decide whether you should go univentricular or biventricular, and a too-aggressive approach in this group of children may be a little bit dangerous. We are at this moment now studying our own Ross-Konno infants, and we certainly have had several patients we are looking at in retrospect where we are sorry that we did not go the univentricular way. So could you comment on that, please?

Dr Aszyk: 40%, not 60%, of our patients are still on medical therapy, cardiac medical therapy, of whom one is on ACE inhibitors and diuretics due to renal failure, not to heart failure. The left ventricular outcomes in our echo follow-up actually are pretty good. We had shortening fraction of 40% on average, and the function of the autograft was excellent. We did not see any diastolic restriction, diastolic impairment.

Dr Hazekamp: And there were no patients at last follow-up who had pulmonary hypertension whatsoever?

Dr Aszyk: There is one patient with pulmonary hypertension. She also had mitral valve problems, and had already developed pulmonary hypertension. Medical therapy to reduce pulmonary vascular resistance has been started, and at current follow-up she is New York Heart Association class II and already listed for transplant.

Dr V. Hraska: In this patient study, we always did the Konno incision to enlarge the very stenotic aortic valve is removed and the subaortic fibrotic area excised, I only need to do a partial septal myectomy within the LVOT onto which we can put the autograft. And, in a way, that would preserve the integrity of the right ventricle and the ventricular septum.

Dr Aszyk: In this patient study, we always did the Konno incision to enlarge the left ventricular outflow tract.
outcome at a median follow-up of 6.2 years, especially for infants with no structural abnormality of the mitral valve. Aortic regurgitation was absent or trivial in all cases, with complete and sustained resolution of LVOTO, as well as preservation of ventricular function. This outcome is concordant with my personal Konno-Ross operation experience in neonates and infants operated over the past 15 years in the USA and Australia. My impression is that the experience (published and unpublished) of other groups would largely support the data presented herein [2-7]. There are a few comments and observations that I would like to offer regarding the Konno-Ross operation in infancy:

(i) The name Konno-Ross operation itself may be misleading as this operation bears little technical resemblance to the Konno operation as initially described [8]. A pulmonary autograft in a small baby virtually always requires an annular and LVOT enlargement, but seldom requires prosthetic patch material [9].

(ii) Problems inherent in older patients (especially young adults) undergoing the Ross operation, such as root dilation and insidious acquisition of aortic insufficiency, may not be as prevalent in neonates and small children undergoing Konno-Ross operation [1, 5, 7, 10]. In many ways, the neonates constitute quite a different cohort.

(iii) The comparison group for the outcome of the neonatal Konno-Ross operation should comprise infants with biventricular hearts treated with competing strategies (interventional approaches, surgical LVOT resection and/or aortic valve reconstruction, Norwood procedure, Yasui procedure, Konno operation, Clarke operation etc.). It would be fair to say that all of these approaches have their own technical limitations, early morbidities and somewhat restricted long-term survival free of LVOT or valve reintervention.

(iv) Detractors of the Ross strategy point out repeatedly that the Ross operation potentially creates two-valve disease, and the Ross-Konno operation with mitral plasty threec valve disease. Clinical experience and current long-term data suggest that even if this were the case, on balance the outcome is superior to that achieved with less definitive solutions in similar patients.

(v) Procrastination in applying the Ross-Konno solution (in favour of multiple surgical or interventional palliations) may lead to hypertrophy and LV diastolic dysfunction that can compromise results of the definitive surgery. Likewise, creation of aortic insufficiency with balloon valvotomy in a neonate may compel the surgical team to perform an extensive surgical procedure under non-ideal conditions.

(vi) The Univentricular Survival Advantage Score proposed by Hickey et al. has been useful in decision-making regarding the Konno-Ross operation for patients with borderline left ventricles, but as noted by Oka et al., the bias introduced by mitral abnormalities requiring surgical treatment may not be completely accounted for [3, 10]. This type of decision-making therefore needs further refinement.

(vii) The technical aspects of the Konno-Ross operation have been simplified and standardized over the past decade, and I would consider it to be both teachable and reproducible. Certainly the surgical skills needed for the arterial switch, aortic root translocation etc. are similar to those required for the neonatal Konno-Ross operation. The commonly associated lesions (arch obstruction, mitral disease, endocardial fibroelastosis (EFE), multiple ventricular septal defects) do increase the complexity level, and it is therefore the neonates who remain at greatest risk. Application of other strategies does not necessarily address this problem.

The immediate benefit of the Konno-Ross operation strategy in the anatomically suitable and critically ill neonate is undisputed, as competing strategies are neither as effective nor safer. The real question centres on expanding the indications to offer primary Konno-Ross operation to young infants who could be well palliated in the short term with other strategies. The favourable longer-term follow-up data presented by the Sankt Augustin group support this concept, and once again point out the continued challenge of mitral disease and EFE and their association.

REFERENCES