January 2002 to December 2011 were included: 1258 (59%) in-born neonates whose delivery was planned in our institution, 799 (38%) terminations of pregnancy, and 73 (3%) foetal deaths. For in-born, planned delivery was classified as ‘certainly justified’ for 899 (71%) for the following reasons: Rashkind atrioseptotomy in 344 cases, risk of aortic coarctation in 272 cases, ductal patency needed for pulmonary flow in 107 cases, ductal patency needed for systemic flow in 93 cases, need for an immediate intervention in 83 cases. For the remaining 359 in-born, planned delivery was classified as ‘potentially justified’ for the following reasons: possible need for ductal patency for pulmonary flow in 156 cases, for systemic flow in 35 cases (3%). Incomplete congenital heart disease diagnosis in 94 cases, need to monitor neonatal tolerance of the defect in 51 cases. In these 359 in-born at risk, rationale for planned delivery was reviewed after birth. A posteriori, it was not necessary for 249 in-born (20%) in whom no intervention was needed during the first week, and confirmed to be necessary for 110 in-born (9%) – 32 in whom diagnosis was different with a direct influence on management and and with 78 who needed an intervention during the first week. 

Conclusions.— Our study demonstrates that only one fifth of foetal congenital heart diseases delivered in a tertiary reference centre appears to be unnecessary. Conversely, one third of in-borns with only possible post-natal risk of cardiac complication were appropriately delivered in our institution, as they needed immediate specialized management.

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06 Long-term follow-up after heart transplantation in very young children
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Heart transplantation in young children and infants may be controversial. The aim of this study was to review long-term follow-up of heart transplanted small children and assess prognosis and outcomes.

Material and methods.— Patients who underwent orthotopic heart transplantation (OHT) within the first 3 years of life were included in the study. Demographics, clinical data, events, outcomes and survival were assessed.

Results.— Among 96 paediatric heart transplantations performed in a French single-centre, 25 patients who underwent OHT at ≤3 years of age, were included in the study (10 males, 15 females). Among them, 10 (40%) were on VAD support at the time of OHT. Age at OHT was 1.5±0.9 years (median 1.2). Underlying cardiac disease was congenital in four (16%) or idiopathic cardiomyopathy in 21 (84%). Post-transplant follow-up was 7.1±7 years (range 1day to 22.7) and was >10years in seven cases (28%). Three patients died at first day, second year and fourth year post-transplant.

Mean age of survivors at the time of the study was 9.1±7.3 years (range 1.5 to 23.6). One acute rejection episode occurred at first month post-transplant and one at 11th year. One patient had post-transplant lympho-proliferative disease at 14th year post-transplant and was successfully cured. Graft coronary disease occurred in two cases (8%), who underwent second heart and kidney transplantation at 16th and 22nd year after first transplant. All other cases were free from coronary disease with normal graft function. End-stage renal failure occurred in the two re-transplanted cases. Significant severe renal dysfunction was present in three cases (no dialysis), moderate in three cases, and 17 had normal renal function. Linear growth ranged within normal in all patients, except the two cases with end-stage renal failure, despite continuous low dose steroid therapy in 80% of the survivors. All patients are in NYHA class I, except the two re-transplanted cases who were in NYHA class IV at the time of second transplant. Patient survival was 96% at 1-year, 90.7% at 3-year and 83% at 10-year post-transplant follow-up. Graft survival was respectively 96%, 90.7%, 83% and 66% at 1-, 3-, 10- and 16-year follow-up.

Conclusion.— Long-term survival of very young heart transplant recipient is fairly good, with a low incidence of graft coronary disease and optimal functional status and growth.

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07 Anatomy of the ventricular septal defect in congenital heart defects: Random or systemic association?
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Introduction.— A ventricular septal defect (VSD) is part of most congenital heart defects (CHD).

Aim of the study.— To determine the distribution of the anatomic types of VSD in CHD.

Material and methods.— We analyzed morphologically 1178 heart specimens with CHD from the anatomic collection of the French Reference Center for Complex CHD. Special attention was paid to the localization of the VSD: muscular, membranous, outlet located between the two limbs of the septal band, inlet. The specimens were classified according to the anatomic and clinical classification of CHD (ACC-CHD).

Results.— A VSD was present in 67% of all hearts and was:
—constant, of a single type, in tetralogy of Fallot and variants and common arterial trunk: outlet, complete atrioventricular canal (CAVC): inlet, and double-inlet left ventricle (DILV): muscular;
—not constant with a predominant type, in 96% of discordance (inlet 82%), 62% of heterotaxy syndromes (Htx, inlet 93%) 93% of interrupted aortic arch (outlet 80%), 87% of double outlet right ventricle (outlet 77%);
—not constant, of variable type, in 68% of aortic coarctation (CoA: outlet 44%, membranous 35%, muscular 21%), 34% of transposition of the great arteries (TGA: outlet 40%, membranous 25%, muscular 25%, inlet 10%);
—rare, in anomalies of pulmonary veins (5%), Ebstein anomaly (14%), double-inlet right ventricle (10%), coronary anomalies (25%);
—isolated in 10% of all VSD: outlet 44%, membranous 36%, muscular 18%, inlet 2%.

Associations were:
—outlet VSD: 60% ‘‘conotruncal’’ defects (CTD), 10% TGA;
inlet: 57% CAVC, 13% DD, 10% Htx;
muscular: 33% DILV, 26% TGA, 13% isolated;
membranous: 30% TGA, 28% isolated, 16% CoA.

Conclusion.— The VSD is an integral part of the phenotype in some CHD (CTD, CAVC, and DILV). In CoA and TGA the VSD is not constant and its anatomic distribution is similar to that in isolated VSD, indicating a likely random association. This reinforces the hypothesis of different genetic mechanisms in TGA and CTD. This original approach, using the anatomic characteristics of one part of the phenotype, could provide new insights in the grouping and aetiology of CHD.

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