

## Univentricular Hearts in Denmark 1977-2009: Incidence and Survival

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### Introduction:

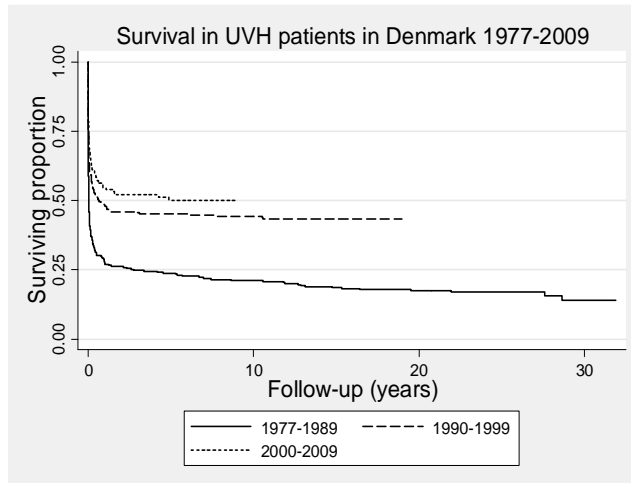
Over the past four decades outcome has improved in children born with a functionally univentricular heart (UVH). This study describes changes in incidence and survival among UVH patients born in Denmark from 1977 to 2009.

### Methods:

Using nationwide registries we identified UVH patients and induced abortions due to UVH in the fetus. The diagnosis was confirmed through local surgical and fetal ultrasound registers, the national register of autopsy, and by review of medical and echocardiography reports. If UVH could not be confirmed the patient was excluded.

### Results:

822 cases were identified. The incidence of UVH (including induced abortions) was evenly distributed in the period (mean 0.400 per 1000 births, SD 0.056). Incidence of liveborn UVH patients however showed a downward tendency in the period 2000-2009. Outcome of birth groups are shown below:



Birth group	Born with UVH, n	Abortion, n	Alive 2009, n	Fontan/TCPC, n (dead)	Type of Fontan, n (dead)		
					1	2	3
1977-89	277	0	43	42(10)	13(4)	19(6)	10(0)
1990-99	248	6	106	99(4)	1(0)	66(4)	32(0)
2000-09	191	100	99	74(4)	0	5(2)	69(2)

Fontan1: Atriopulmonary connection; Fontan2: Lateral tunnel; Fontan3: Extracardiac tunnel.

The Kaplan-Meier curves above (abortions excluded) show an increase in survival between 1977-89 and 1990-99 (log-rank,  $P < 0.0001$ ) and between 1990-99 and 2000-09 (although not statistically significant, log-rank,  $P = 0.20$ ). Survival and numbers of nondeceased/noncensored cases are shown in the table below.

Birth group	n	1 year survival (95% CI)		5 year survival (95% CI)		10 year survival (95% CI)		20 year survival (95% CI)		30 year survival (95% CI)	
		Survival	n	Survival	n	Survival	n	Survival	n	Survival	n
1977-1989	277	0.28 (0.23-0.33)	76	0.24 (0.19-0.29)	65	0.21 (0.17-0.26)	57	0.17 (0.13-0.22)	45	0.14 (0.09-0.20)	6
1990-1999	248	0.48 (0.42-0.54)	121	0.45 (0.39-0.51)	113	0.44 (0.38-0.50)	96				
2000-2009	191	0.54 (0.47-0.61)	93	0.50 (0.43-0.57)	49						

### Conclusions:

The incidence of UVH was constant throughout the period indicating constant detection rate over time. The incidence of liveborn UVH patients was falling after 2000 due to introduction of fetal ultrasound. Survival has improved from the 1980ies but has not significantly improved in the last 20 years.