

The Swedish Registry on Grown-Up Congenital Heart Disease



Annual Report 2007

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PUBLICATIONS

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INFORMATION about Swedish GUCH-services and links:

www.guch.nu

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AIMS & BACKGROUND

The number of grown-up patients with congenital heart disease is steadily increasing due to the success story of paediatric cardiology and cardiac surgery. In Sweden, there are approximately 20-25.000 adults with a congenital cardiac lesion, all degrees of severity included. The patient population of adults with congenital heart disease is now outnumbering that of children. Children who in past times died during childhood now reach adult life and caregivers encounter patient groups that have not existed before. Knowledge and experience are therefore limited.

The incidence of congenital heart disease (CHD) in live-born children is around 0,8 % and depending on the birth rate there is a yearly influx of 750-1000 children with CHD in Sweden. In many of the patients with complex heart malformations, operated upon in childhood, the operation is not curative but corrective or even palliative. These patients will need advanced medical care and will demand both medical and surgical treatment later in life. The long-term prognosis is often unknown. The wide spectrum of different malformations as well as the continuous change and improvement of the surgical and medical treatments makes it difficult to obtain a clear picture of the long-term outcome.

The last decade's percutaneous catheter techniques have developed into an alternative to cardiac surgery. The post-operative pain is less and the recovery time much faster. However, the medium- and long-term results of these techniques are still very limited.

Heart disease also influences social factors such as the professional career, raising a family and the choice of leisure-time activities. A question of great importance for many women with heart disease is their ability to become a mother. Since patients with complex CHD previously rarely reached adulthood the experience of pregnancies in this group of patients is limited.

Even patients with so called simple lesions have a need of comprehensive information and assessment by a specialist. It is therefore desirable that all patients reaching adult age and not completely cured during childhood should have the opportunity to meet a cardiologist specialized in Grown-Up Congenital Heart disease (GUCH).

There is an ongoing project to create a Swedish registry covering congenital heart disease in all ages and the associated cardiac surgery (SWEDCON). It will start running in late 2008 and the present Swedish registry on adult congenital heart disease will be a part of that.

The aims of the Swedish registry on adult congenital heart disease are:

- To assure the quality of the medical care for grown-up patients with congenital heart disease
- To create conditions for national guidelines
- To give feed-back to paediatric cardiology and heart surgery by long-term follow-up
- To identify patients with a high risk for late complications
- To create conditions for national and international research
- To serve as a source of information when counselling in the individual case

COVERAGE

To be included in the registry the patient has:

- To have been assessed at least once at one of the 7 national GUCH centres (= those participating in the registry)
- To have a congenital structural cardiac malformation
- To be ≥ 16 years of age

Patients with simple lesions that do not need specialized care are therefore not included in the registry. However, when such a patient suffers a complication referral to a specialized unit often takes place and consequently registration occurs. There is awareness that this bias may suggest a darker prognosis of simple lesions than it really is. However, there is a growing interest from lower levels of care to participate in the registry thereby improving the coverage of uncomplicated simple lesions. Furthermore, in the future integrated registry (SWEDCON) including all ages, the paediatric part will include all Swedish patients with a congenital cardiac lesion. In patients with complex CHD there is a well organised transition from paediatric cardiology to specialized GUCH centres and the coverage of the registry is by and large complete. The same is true for patients undergoing cardiac surgery or catheter interventions as such procedures are made only at GUCH centres.

SVENSKA HJÄRT-KÄRLREGISTREN (SWEDISH CARDIO-VASCULAR REGISTRIES)

The Swedish registry on grown-up congenital heart disease, as well as the future SWEDCON registry, is a part of Svenska Hjärt-kärlregistren (Swedish Cardio-vascular registries) which is an association of several cardio-vascular registries using the technical platform and the experience of Uppsala Clinical Research Center (UCR) lead by Professor Lars Wallentin.

DATA COLLECTION & FEED-BACK

The registry, founded in 1992 initially included two centres. Since 1998 it comprises all the 7 regional GUCH centres in Sweden. Since 2005 the input of data is through the Internet to a technical platform and server provided by the Uppsala Clinical Research Center. Updated registry data and analyses are immediately obtained by the Internet as:

- Annual reports (public)
- Preformed descriptive and analytical reports which are daily up-dated for users. National data and data from the user's own centre are obtained.
- Export of data to excel or statistical software when issues that are not covered by the preformed analyses occur.

Registry users regularly meet to discuss technical and analytical issues in order to **improve the quality of care** of Grown-Up Congenital Heart disease. The registry also has a very important **descriptive function** as it deals with groups of patients that have not existed before. Important information can easily be reached and used in an individual case.

The registry has promoted the cooperation between the Swedish GUCH centres and has contributed to a rather homogeneous opinion in the profession about the care of adults with congenital heart disease in Sweden. In an international perspective the registry is unique.

QUALITY AND VALIDITY

The registry has functions for control of missing or incorrect data and for unusual combinations. The on-line analytical possibilities allow identification and checking of outliers.

The registry contains data on the type(s) of underlying cardiac condition and performed interventions, surgical or catheter-based. Detailed information about medication, present condition, and echocardiographic data are gathered at every visit. The registry is longitudinal, the aim is to follow the patient until the end of life. Simple social variables and the outcome of pregnancy are recorded. Every time a visit, investigation or intervention is made information is updated to give the opportunity to get data for long-term follow up.

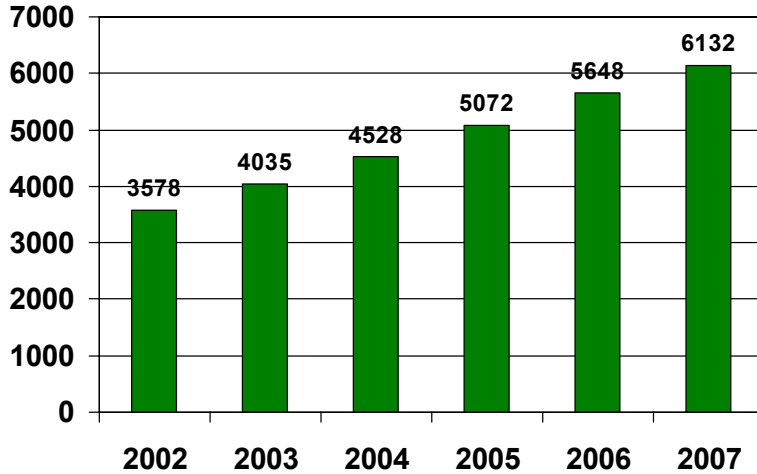
Since 2005 quality of life assessment in terms of the EQ-5D instrument has been added. For catheter interventions there is a structured follow up with emphasis on results and complications.

Control of validity is made by random comparison of registry data with original data (for example in hospital reports).

REGISTRY DATA

TOTAL NUMBER OF PATIENTS IN THE REGISTRY

2002-2007

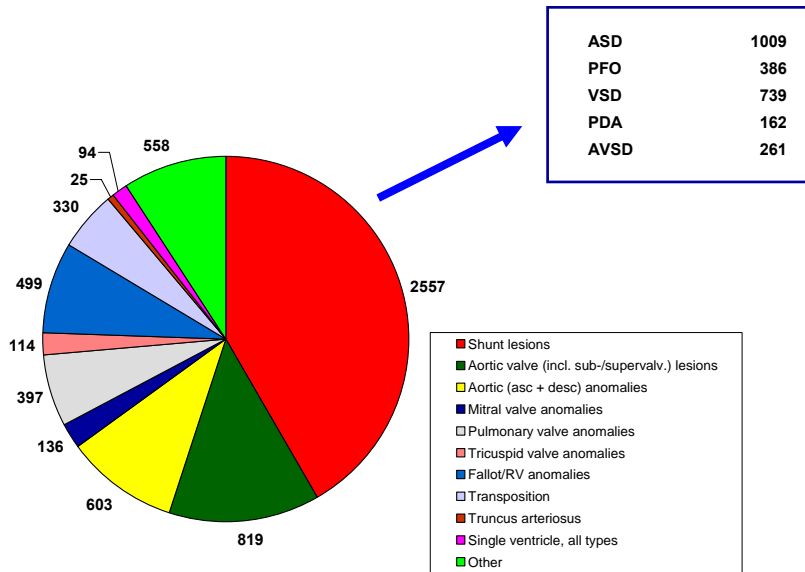


For a long period of time there has been a yearly increase of about 500 patients and the total number of patients in the registry is now exceeding 6000. Thus, around 25 % of the estimated number of all adult patients in Sweden with a history of congenital heart disease have been evaluated at specialized GUCH centre. Internationally it has been suggested that this figure should be 25-50 % suggesting shortcomings of the Swedish organisation in the care of grown-up congenital heart disease. There has been a fear that there is a loss of patients when transitioned from paediatric cardiology to GUCH services due to suboptimal referral routines. The development of an integrated paediatric-adult registry, as mentioned before, will clarify this and help to improve the transition.

Age- and sexdistribution			
		Females	Males
N		3020	3112
Age, years	Mean	39	36
	Median	35	32
	Range	16-87	16-92

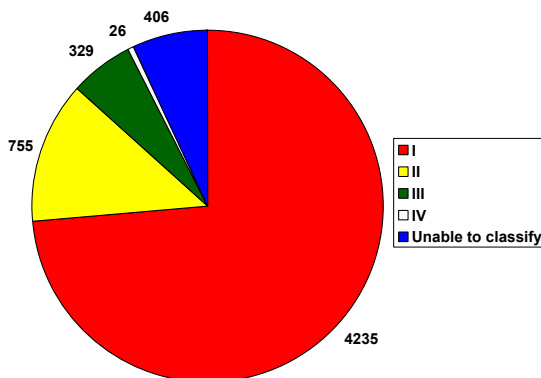
More than half of the patients are younger than 35 years of age. Being in the early phases of adult life, their needs may be very different from that of other types of heart disease which usually are found in much older patients.

DISTRIBUTION OF MAIN DIAGNOSES



Shunt lesions constitute 1/3, atrial septal defect (ASD) making up 17% of all patients. In paediatric cardiology the corresponding figure for ASD is 10%. There are several possible reasons for this discrepancy, one is that ASD to a large extent is diagnosed in adult life because of subtle symptoms and signs during childhood. If so, there is a feed-back message to paediatric cardiology. Patients with patent foramen ovale (PFO) are by and large patients with a catheter intervention because of presumed paradoxical embolism, as the mere presence of an isolated PFO does not merit inclusion in the registry. During the last five years the number of patients with “single ventricle” – a group with extensive demand for advanced care - has increased from 36 to 94.

NYHA FUNCTIONAL CLASSIFICATION at latest appointment



Functional assessment according to the NYHA (New York Heart Association) functional classification is regularly made and changes over time can be followed as well as comparisons between different groups of patients. Nearly 3/4 of the patients regard themselves

as asymptomatic. Although the distribution of functional classes has not changed during recent years, the absolute number of highly symptomatic patients (NYHA III and IV) has increased, by around 35 % the last five years, due to the general increase of patients. As such patients tend to have a high demand for care, in the out-patient clinic as well as in the in-hospital setting, these increasing numbers are important to consider when allocating resources to the GUCH services.

NYHA related to main diagnosis

Percentage of patients (still alive) in NYHA III or IV at the latest appointment	
	<u>%</u>
Aortic (asc + desc) anomalies	2
Aortic valve (incl. sub-/supervalv.) lesions	2
Single ventricle	19
Fallot/RV-anomalies	7
Mitral valve anomalies	3
Pulmonary valve anomalies	4
Shunt lesions	4
Transposition	6
Tricuspid valve anomalies	6
Truncus arteriosus	22
Other	4

Severe functional impairment (NYHA III- IV) is common in complex lesions like single ventricle and truncus arteriosus, and not uncommon in tetralogy of Fallot and transposition. Compared with the period 2004-2006 the proportion of highly symptomatic patients in the two former diagnostic groups seems unchanged while it has slightly decreased in tetralogy of Fallot and transposition.

QUALITY OF LIFE ASSESSMENT

EQ-5D

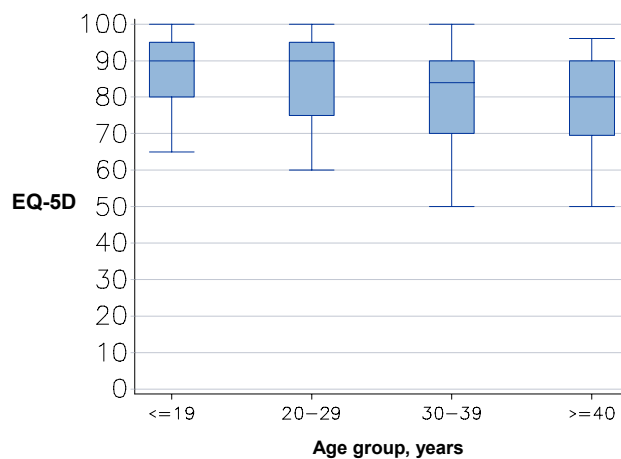
<u>NYHA</u>	"Simple" lesion	"Complex" lesion	N Simple/complex
I	83	85	1578/324
II	70	73	267/137
III	58	58	90/50
IV	11	15	4/1
Not able to classify/unknown	73	72	212/40

Quality of life at latest doctor’s appointment. The EQ-5D rating related to NYHA functional class in patients with complex lesions (=main diagnosis is Fallot, transposition, single ventricle or truncus arteriosus) and simple lesions (= not complex lesions).

When the registry was web-based during 2005 life quality assessment by means of EQ-5D was added. The EQ-5D instrument consists of 5 questions regarding general aspects of life and a ruler graded from 0 to 100 (“life quality thermometer”). The patient is asked to mark on the ruler the present level of life quality where 0 represents “worst possible” and 100 means “best possible”.

It seems obvious that life quality as measured here is associated with the functional class but not with the complexity of the underlying cardiac malformation. When time goes it will be possible to analyse to what extent given care and therapy may influence quality of life.

EQ-5D related to age



Quality of life by EQ-5D related to age groups (N=1434). Age does not seem to significantly influence life quality.

MORTALITY

Premature death due to cardiac reasons is not infrequent. Among the 6132 registered patients there were a total of 294 deaths (4,8%) at the end of 2007 and 34 of them occurred during 2007. As the registered patients regularly are checked against national vital statistics, deaths unknown to the participating centres will be included. However, in those cases, the cause of death is not automatically transformed to the registry and this explains the substantial proportion of deaths of unknown cause. This must be improved in order to obtain more accurate statistics. The relatively high age at death in the “unknown” group may indicate that age-related acquired disease is of importance. The pattern of mortality does not seem to have changed during recent years.

	N	Age at death, years	
		Mean	Range
Mortality, total	294		
by which suddenly	90		
by which perioperatively	7		
Cardiac death	143	44.5	24-87
Non-cardiac death	55	55.1	18-85
Unknown	96	62.2	22-93

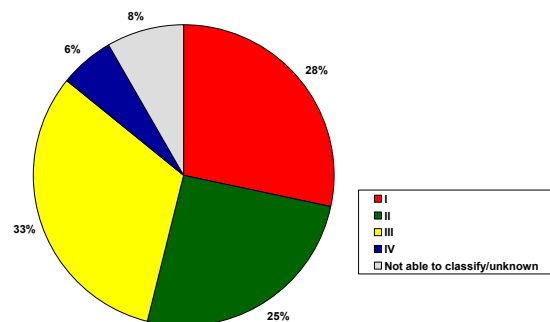
Cardiac death dominates. Half of the deaths are reported being cardiac, however, this is likely to be an underestimation due to the rather large number of deaths of unknown cause.

At least 25% of the deaths are sudden which underlines the importance of arrhythmia as a common and serious complication in adult congenital heart disease. This emphasises the need to improve diagnostic and prognostics tools as well as treatment strategies to reduce the incidence of life-threatening arrhythmias.

In more than 50 % of the deaths there were no or only mild symptoms at the closest appointment preceding death.

A plausible explanation would be that lethal arrhythmias also occur in patients who seem to do well. Again, the need to find more accurate prognostic tools for arrhythmia is apparent .

Mortality related to NYHA classification at the latest appointment before death



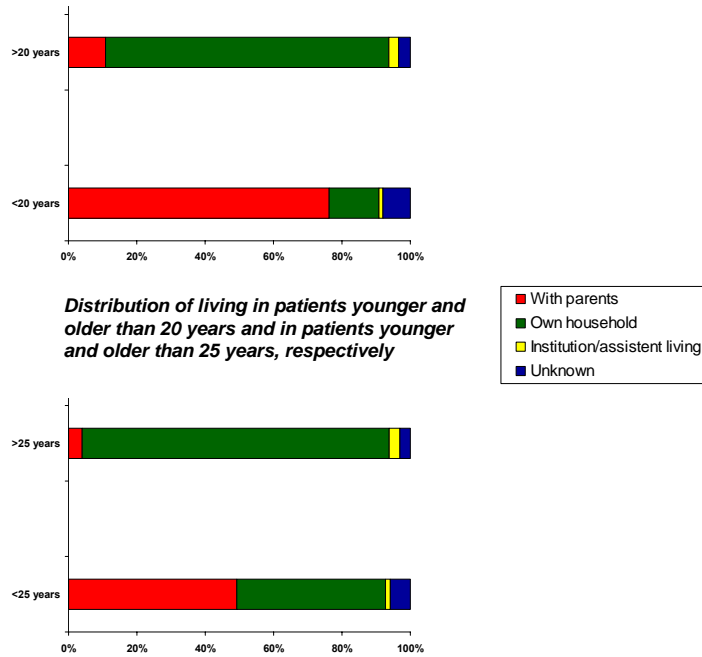
Mortality and age at death related to main diagnosis

<u>Main diagnosis</u>	<u>Total number</u>	<u>Mortality</u>		<u>Mean age, years</u>	
		N	% of total	at death	in those alive and seen during 2007
Shunt lesions	2557	141	6	60.3	43.2
Aortic valve (incl. sub-/supervalv.) lesions	819	20	2	46.8	33.0
Aortic (asc + desc) anomalies	603	10	2	51.9	37.3
Mitral valve anomalies	136	3	2	36.3	29.8
Pulmonary valve anomalies	397	17	4	43.6	35.5
Tricuspid valve anomalies	114	6	5	62.8	40.0
Fallot/RV anomalies	499	25	5	52.6	34.5
Transposition	330	23	7	36.1	29.0
Truncus arteriosus	25	3	12	35.7	31.7
Single ventricle, all types	94	15	16	31.9	27.9
Other	558	28	5	45.5	36.3

Complex lesions like single ventricle, truncus arteriosus and transposition carry a higher mortality risk than many of the "simple" lesions in spite of a younger age profile. It is to be noted that a prerequisite for registration is that the patient has reached adult life (≥ 16 years of age) and the given figures do not consider mortality during childhood. In shunt lesions and tricuspid valve disease the mean age at death is around 60 years, suggesting that acquired disease and "natural" causes may have played a role.

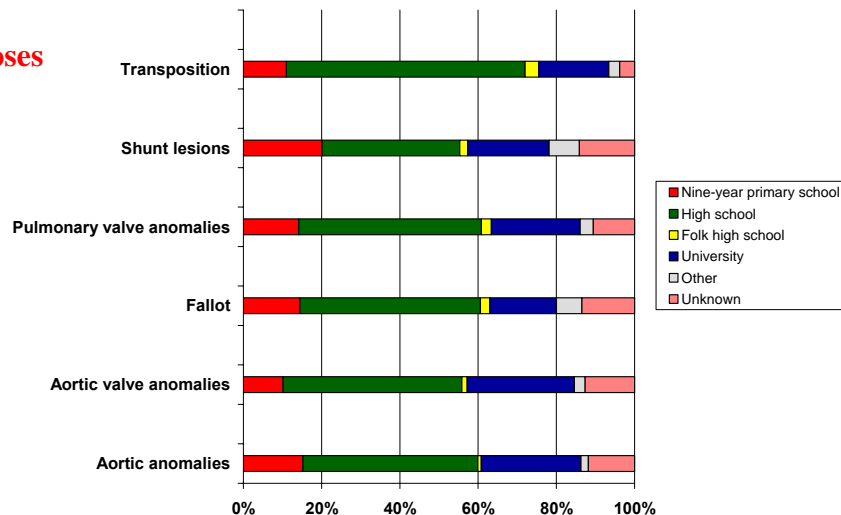
SOCIAL VARIABLES

Living



There is a fear that parental overprotection would hamper normal development of adult independence and autonomy. Having a household of your own could serve as a marker of social independence. Among patients older than 20 years of age 11% live with their parents and in those older than 25 years of age this is 4%. Two years ago the corresponding figures were 12% and 4% respectively. Thus, expressed in this way the autonomy process of grown-up congenital heart disease patients does not seem to be very different from what is normally seen. This is an important feed-back to paediatric cardiology.

Education per diagnoses



The highest level of education based on the main diagnosis. In general the educational level is high. Compared with the period 2004-2006 the pattern has not significantly changed. For a more detailed analysis the different age structures in the different groups must be considered. "Shunt lesions" contains a large proportion of middle-aged and older patients and the data may reflect the standard of education in former times. Patients with transposition tend to be young and many of them have not yet reached the age for university studies.

TRANSPOSITIONS**Heart failure therapy in patients with congenitally corrected transposition (CCTGA) and Mustard/Senning repaired transposition related to systemic ventricular systolic function and NYHA functional class.**


<u>Systemic ventricular systolic function</u> (by echocardiography)	N
Normal	88
Mild dysfunction	65
Moderate dysfunction	75
Severe dysfunction	20
Σ =	248
<hr/>	
Patients with moderate or severe systemic ventricular dysfunction	95
<i>In those:</i>	
No medical therapy for heart failure, N	38 (40%)
ACE/AII-antagonist therapy, %	52
<u>Patients in NYHA III or IV</u>	28
<i>In those:</i> No medical therapy for heart failure	7(25%)

In congenitally corrected transposition and in Mustard/Senning repaired transposition a morphologically right ventricle serves the systemic circulation. Echocardiographic assessment of the systemic systolic ventricular function, mainly by “eye-balling” is reported in 248 such patients. Severe dysfunction could be interpreted as an ejection fraction (EF) around 30% or less. Moderate dysfunction would correspond to an EF in the range 30-45%, and mild dysfunction to EF 45-55%.

40% of those with a severe or moderate systolic dysfunction of the systemic ventricle at the latest exam, did not receive medical heart failure therapy (= any of ACE-inhibitors, AII-antagonists, diuretics, beta-blockers or digoxin). About half of them were treated with an ACE-inhibitor or an AII-antagonist. In those with marked functional impairment (NYHA III and IV) appropriate therapy was absent in 25%, suggesting a low standard of care. However, the reason for this is unknown as the registry does not give information on the basis for caregiver’s decisions or patient factors for example compliance.

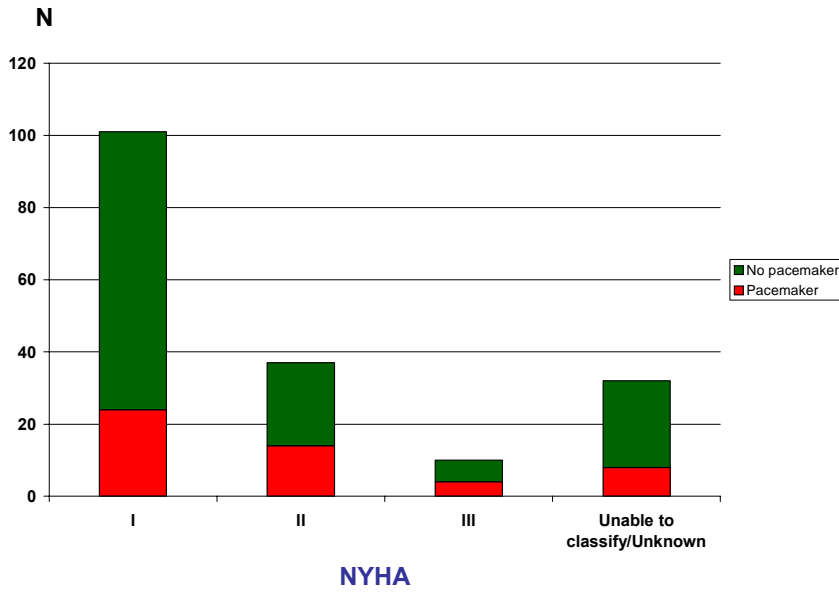
Mustard/Senning repaired transposition; Heart failure therapy related to systemic ventricular systolic function and NYHA functional class during the period 2002 – 2007.

Systemic ventricular systolic function (by echocardiography)						
Normal				63	57	49
Mild dysfunction				16	31	42
Moderate dysfunction				48	49	54
Severe dysfunction				8	10	13
			Σ =	135	147	158
	2002	2003	2004	2005	2006	2007
Patients with moderate or severe systemic ventricular dysfunction, N	41	47	51	56	59	67
<i>In those:</i>						
<i>No medical therapy for heart failure, N (%)</i>	27(66%)	31(66%)	37(73%)	35(63%)	28(47%)	27(40%)
<i>ACE/AII-antagonist therapy, %</i>	27%	26%	20%	30%	42%	51%
Patients in NYHA III or IV*, N (% of all)				7(6%)	10(7%)	9(6%)
<i>In those: ACE/AII-antagonist therapy, %</i>				14%	50%	67%
				<small>*No patient in NYHA IV</small>		

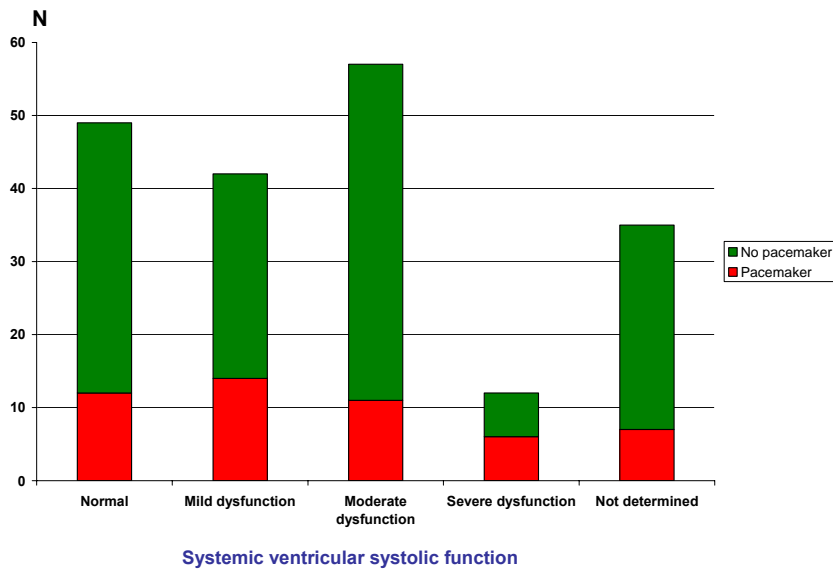


More than 40 % of the transpositions with a Mustard/Senning repair have systemic ventricular systolic dysfunction of moderate degree or worse. During recent years there has been a gradual increase in the use of ACE-inhibitors/AII-antagonists in such patients and the proportion of patients without any medical heart failure therapy (= any of ACE-inhibitors, AII-antagonists, diuretics, beta-blockers or digoxin) has declined. The standard of care, in this aspect, has improved and one reason could be the discussion registry data has stimulated. However, the basis for ACE-inhibitor and AII-antagonist treatment is transferred from experience obtained in acquired heart disease and it is important to know that scientific evidence for treating asymptomatic ventricular dysfunction in the Mustard/Senning setting is lacking.

Mustard/Senning repaired transposition; Pacemaker therapy related to NYHA functional class and systemic ventricular function



More than a quarter (28%) of all Mustard/Senning patients have a pacemaker. The main reasons are sinus node dysfunction and bradyarrhythmia in combination with tachyarrhythmia where treatment of the latter, often with negative chronotropic drugs, necessitates the insertion of a pacemaker. Reduced physical performance, in terms of NYHA class, is associated with a higher prevalence of pacemaker therapy.



With the exception of those with a severely dysfunctional systemic ventricle the presence of a pacemaker is rather equally distributed in the groups of different ventricular performance.

If reduced physical capacity is linked to a higher risk of atrial tachyarrhythmia which in turn, as mentioned above, could be associated with pacemaker therapy, this would be an explanation. However, scientific reports have demonstrated that systemic ventricular dysfunction in the Senning/Mustard setting is linked to a substantial increased risk of atrial tachyarrhythmias and one would then expect to find a more obvious relationship between the frequency of pacemaker therapy and systolic ventricular function. This was not the case here.

Thus, pacemaker therapy is associated with reduced physical performance while the association with systemic ventricular function is much less prominent. The reasons for this are unclear and it can not be ruled out that pacemaker therapy per se contributes to the functional impairment. Deeper analysis of registry data, not performed in this report, may help to clarify this.

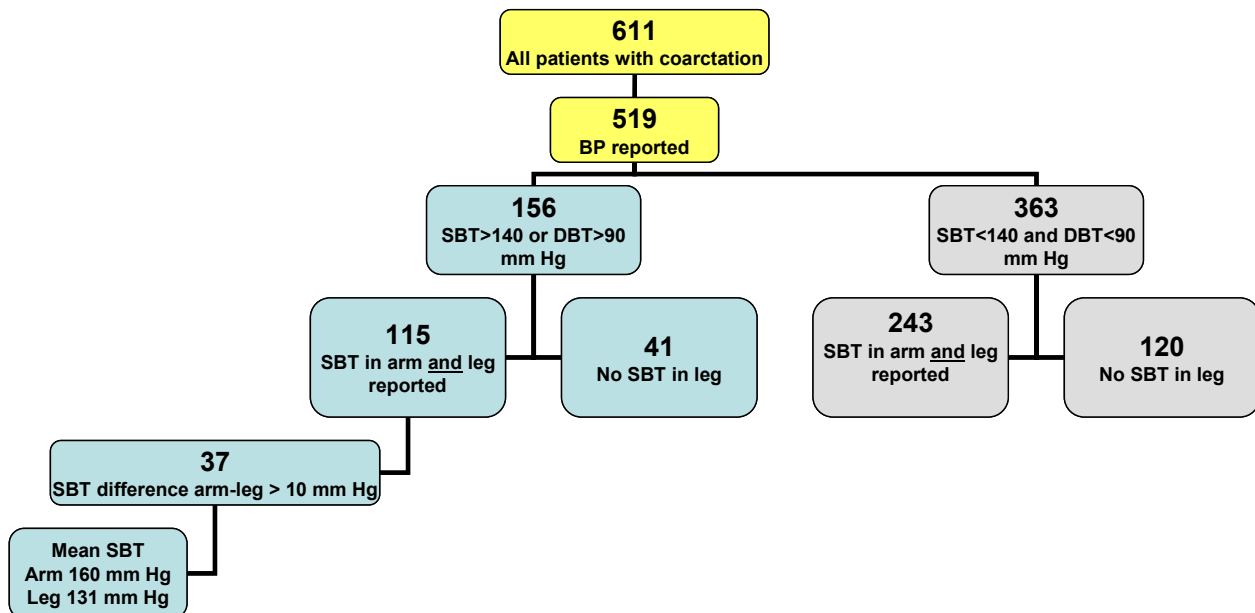
AORTIC COARCTATION

Aortic coarctation, an obstruction usually located in the very distal aortic arch, constitutes more than 6 % of all congenital cardiac malformations. In Sweden that would correspond to a yearly incidence of around 50 cases. Surgical repair of aortic coarctation has been available for more than 60 years. Provided that most of them have survived there would be around 2000 adult Swedes with coarctation. Only 1/3 of them, 611 patients, are included in the registry. 61% of the patients are male.

Aortic coarctation is, incorrectly, regarded as a simple and localised malformation and many patients are followed, if at all, in non-specialised units. However, aortic coarctation is rather a syndrome including aortic valvulopathy (bicuspid valve), ascending aortic dilatation and a risk of hypertension even when the anatomic result of surgery is good. Of the 611 patients with aortic coarctation 79 (13%) have had aortic valve surgery. 24 patients (4%) have dilatation of the ascending aorta as a secondary diagnosis and this should be regarded as a very minimum because cases with a mild widening of the aorta are, for different reasons, not labelled in the diagnostic part of the registry.

With the advent of the integrated paediatric-adult registry (2009) and the extension of the registry to lower-level services it will be possible to get data from a majority of patients with aortic coarctation and get a better idea about the number of patients lost to follow up.

Aortic coarctation; the prevalence of blood pressure measurements in both arm and leg related to blood pressure level



Blood pressure (BP) at the latest appointment. SBT=systolic blood pressure. DBT=diastolic blood pressure. Hypertension is defined SBT>140 or DBT>90 mmHg.

The vast majority of patients have had coarctation repair. Blood pressure is reported in 519 (85%) of the patients. It should have been nearly 100% and routines have to be improved. 156 patients (30%), irrespective if antihypertensive therapy was given or not, were hypertensive at the latest exam. In last year's report it was demonstrated that 2/3 of the coarctation patients with a secondary diagnosis of hypertension did not reach a blood pressure below 140/90 mm Hg at the latest appointment. Although a single blood pressure measurement should not be the sole basis for antihypertensive medication or modification of it, these data suggest a suboptimal care when it comes to hypertension in patients with aortic coarctation.

Blood pressure should routinely be measured both in the upper and the lower part of the body in patients with repaired/unrepaired coarctation. In patients with hypertension this is even more essential as a treatable re-coarctation can be discovered. This has not taken place in around 30% of the patients, irrespective if blood pressure measurements indicate hypertension or not. However, this is a substantial improvement compared to 2005 when the corresponding figure was 62 % and the reason for this is the feed-back of registry data to the working group and the participating centres.

In 32% of those with a blood pressure exceeding 140/90 there was an arm-leg blood pressure difference of 10 mm Hg or more (mean pressure gradient 29 mm Hg) indicating that re-coarctation plays an important role in the development of hypertension in patients with repaired coarctation.

**Surgery and catheter interventions in aortic coarctation 2000 – 2007
(native and re-coarctations)**

	<u>2000 – 2003</u>		<u>2004 – 2007</u>	
	N	Median age, years	N	Median age, years
Catheter intervention	12	22	27	36
Surgical repair	5	37	16	35

During recent years the number of interventions in aortic coarctation has increased. The reason is not clear but one explanation might be a slide in indications. Growing experience and the technical development in catheter treatment such as covered stents have made interventions less risky which would shift the cost-benefit ratio towards active treatment. The introduction of covered stents has expanded the possibilities beyond “re-coarctation” and catheter intervention is now also applied in native coarctation.

CATHETER INTERVENTIONS

Catheter based intervention in congenital heart disease – in paediatrics as well as in adults – has rapidly developed the last decade. The number of catheter interventions in grown-up congenital heart disease has increased manifold. During 2007 there were 181 interventions, the year 2000 there were 41. During the last three years there has been a nearly 50 % increase. Until 2006 catheter interventions were only performed in Gothenburg, Lund and Stockholm. During 2007 Linköping has started a catheter programme, mainly closure of atrial septal defects and patent foramen ovale. The registry has put a special emphasis to watch over the quality of catheter interventions and it is presupposed that reports on results and adverse events regarding the day after, 3-6 months after and one year after the intervention are entered.

Catheter interventions 2005 – 2007 in Swedish GUCH centres

<u>Catheter interventions, numbers</u>			
	All types		
	2007	2006	2005
National	181	159	130
Gothenburg	82	70	69
Stockholm	46	50	31
Lund	30	26	20
Linköping	19	5	2
Uppsala	3	6	8
Umeå	1	2	-
Örebro	-	-	-

Catheter interventions 2007, distribution on types and performing centres

Catheter interventions 2007							
	ASD closure	PFO closure	PDA closure	CoA Stenting	Percutan. valve insertion	EI-phys. ablation	Other
National	65	73	6	14	3	16	4
Gothenburg	31	37	5	5	-	-	4
Stockholm	13	18	1	6	-	8	-
Lund	17	4	-	3	3	3	-
Linköping	4	14	-	-	-	1	-
Uppsala	-	-	-	-	-	3	-
Umeå	-	-	-	-	-	1	-
Örebro	-	-	-	-	-	-	-

ASD=Closure of atrial septal defect. PFO = Closure of patent foramen ovale.
 PDA = closure of persistent ductus arteriosus. CoA = dilatation/stenting of aortic
 coarctation. Other = pulmonary valve balloon dilatation, stenting of pulmonary artery
 obstructions, closure of arterio-venous and coronary fistulae....

A main reason for closing a PFO has been to prevent paradoxical embolisation in patients with a cryptogenic stroke. There is so far no solid scientific evidence justifying routinely closure of a PFO in cryptogenic stroke. This uncertainty gives space for different interpretations and indications for PFO closure which explain the variation in numbers from one centre to another. In a limited number of patients there is a right-to-left shunt causing hypoxemia and then the indication for PFO closure is symptomatic.

Results and complications in catheter-based ASD and PFO-closure during 2007

	<u>ASD</u>	<u>PFO</u>
Total number	65	73
Treatment stopped before attempt	3	0
Treatment failed after attempt	5	3
Treatment completed	57	70
Results		
<u>At 24 hours post procedure</u>		
Complete closure	78%	92%
Trivial residual shunt	13%	7%
<u>At 3-6 months post procedure</u>		
Complete closure	88%	
Trivial residual shunt	8%	
Complications		
Thromboembolism during or shortly after procedure	2	2
Arrhythmia	1	5
Other	1	1
Bail-out surgery	0	0
Perforation/tamponade	0	0
Mortality	0	0

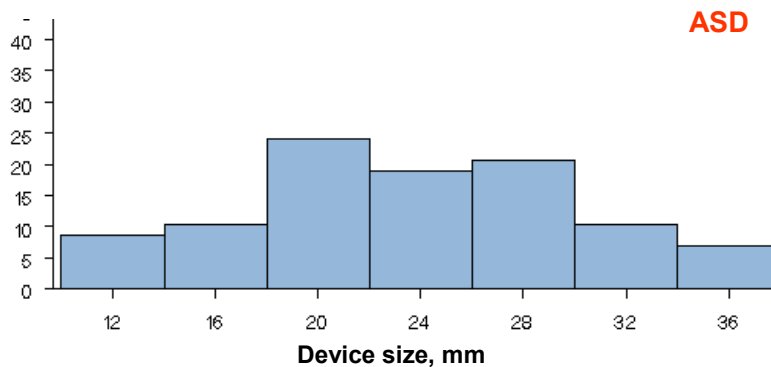
In 12% of the ASD patients and in 4% of the PFO cases the treatment was unsuccessful and a device could not be placed. The anatomical pre-requisites for device closure differ a lot between ASD and PFO. In the ASD group there is a subset of patients with a high surgical risk in whom already from start there are some doubts about the feasibility to close the defect by means of catheter technique but it is attempted. This may explain the higher frequency of unsuccessful procedures in the ASD group.

In ASD the aim is to eliminate the haemodynamic burden of the shunt (“haemodynamic indication”) while in PFO it is to completely close the intra-atrial communication as even small paradoxical emboli can cause harm. Thus, in ASD a successful result would also include a trivial residual shunt. At 24 hours post procedure more than 90% of those with a completed intervention, irrespective if ASD or PFO, have a successful result. At the 3/6 months check-up only 4% of the ASD patients had a suboptimal result and that includes some few patients with multiple fenestrations in whom even a somewhat more than mild residual

shunt had been considered a better option than surgical closure. In the PFO group the numbers as reported at 3/6 months follow-up do not allow a robust analysis.

The number of complications is limited. The reported incidence of thromboembolism (2,7% in the PFO group) is a matter great concern, particularly in the PFO group where the main reason for intervention is to prevent serious paradoxical thromboembolism. However, when analysing PFO closure 2000-2007 the incidence of procedure-related thromboembolism stops at 1,5%. Paroxysmal supraventricular arrhythmia (atrial fibrillation) is a manifestation of the disease in ASD. When supraventricular arrhythmia is associated with PFO closure it is usually transient and develops during or early after the procedure. In the 138 ASD- and PFO-interventions there have been no bail-out surgery, no perforation and no procedure related mortality.

Distribution of used device size in ASD during 2007



Amplatzer® devices are the most frequent type of device. In ASD an Amplatzer® device was used in 90 % of the patients, in PFO 57% of the patients had an Amplatzer® device (33 PFO occluders and 9 ASD occluders). Above is shown the distribution of the device size in ASD, the median size used is 24 mm.

When new types of devices are introduced the registry gives an excellent opportunity for assessment of benefits and shortcomings compared to established brands.

In summary, catheter interventions to close ASD and PFO in Sweden are characterised by high quality with good results and a low incidence of serious complications. However, the medical value of closing PFO in cryptogenic stroke on a routine basis is still an unsolved issue and indications seems to vary between centres.

CARDIAC SURGERY IN GROWN-UP CONGENITAL HEART DISEASE IN SWEDEN 2006 –2007

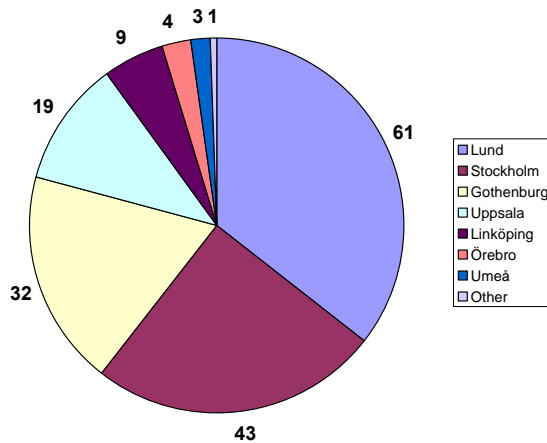
In Sweden, paediatric heart surgery (work load \approx 450 cases yearly) has been centralised to two centres – Lund and Gothenburg – since the early 1990ies. In these two centres tight collaboration between paediatric and general cardiac surgeons have been established in order to provide adult patients with congenital heart disease the experience of both specialities. However, when it comes to surgery of adult patients centralisation has not taken place. There have been reasons and arguments for this, one being that general cardiac surgeons are familiar with particularly aortic and mitral prosthetic valve replacements, another that the treatment of “simple” congenital lesions, like an atrial septal defect, would not be necessary to centralise. However, there is no agreement on the definition of “simple”.

The registry notes the types and the date of cardiac surgery and where it is performed. To be considered “adult” the patient must be 16 years of age or older at the time of the operation. The registry does not provide detailed information about surgical complications in terms of morbidity or mortality, although vital statistics and additional secondary diagnoses can be filled in and then related to the date of surgery when analysed.

During 2006 and 2007 a total of 172 adult patients with congenital heart disease had cardiac surgery (pacemaker and ICD insertion not included). This figure corresponds rather well to what is reported in the Swedish Registry on Cardiac Surgery and it is therefore unlikely that an important number of patients would be missing because of flaws in registration. The adult part of “congenital heart surgery” constitutes around 15% of the total work load.

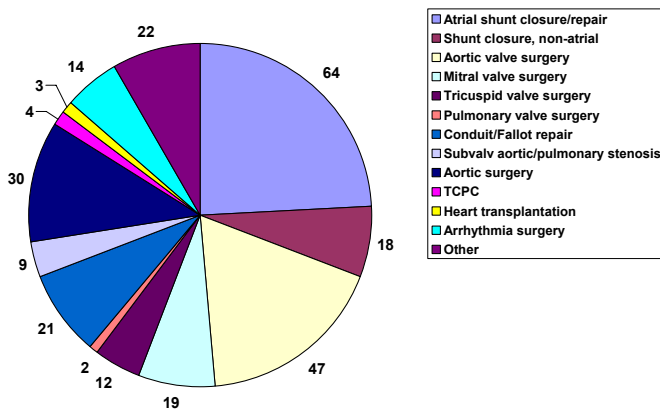
As an operation may consist of more than one surgical measure the corresponding number of measures was 265. The mean and median age at surgery were 41,5 and 40,9 years respectively (range 16-81 years).

Cardiac surgery 2006 - 2007 (n=172); distribution of performing centres

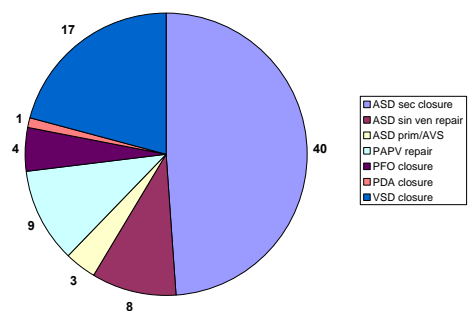


The two centres for paediatric heart surgery – Lund and Gothenburg – perform 54% of all operations. Interestingly, in spite of having about the same number of registered patients the surgical activity in Lund is twice that of Gothenburg, while the opposite is true for catheter interventions. If this is due to preferences or differences in case mix and referral patterns is not known and demands a more detailed analysis.

Cardiac surgical measures 2006 - 2007 (n=265); distribution of types



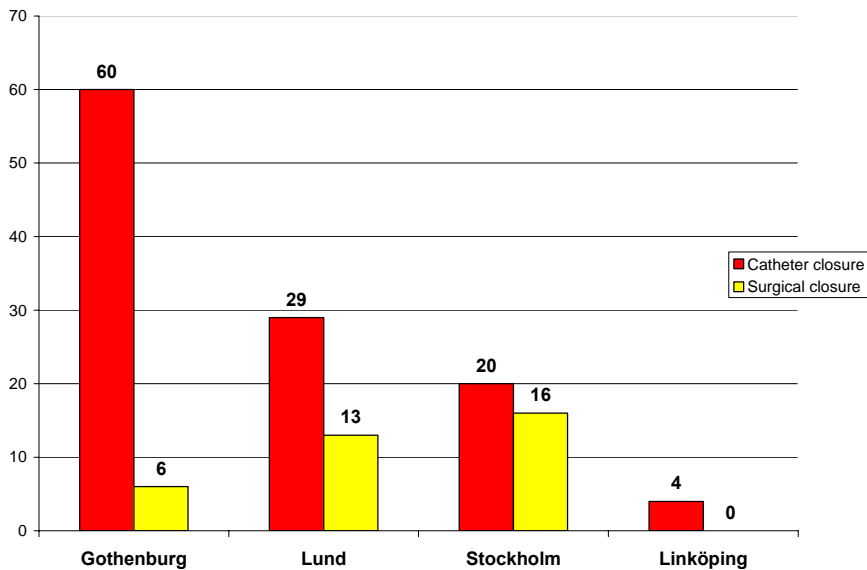
Shunt lesions (n=82), types



1/4 of all surgical measures are aortic or mitral valve surgery; valve replacement or valvuloplasty. In aortic valve surgery it may be isolated or a part of composite surgery for ascending aortic surgery. “Mitral valve surgery” also includes surgical intervention of the systemic AV-valve in transpositions. More than 25% of the procedures constitute closure of a shunt lesion with atrial septal defect (ASD) of the secundum type dominating. However, nearly half of the surgically relieved shunt lesions are atrial lesions of some anatomic complexity and ventricular septal defects (VSD).

TCPC = total cavo pulmonary connection.

Catheter vs. surgical closure of ASD secundum 2006 – 2007 in those centres performing catheter closure



As seen, the proportion between catheter and surgical closure of ASD secundum differ substantially between centres. One reason is the case mix. Linköping started a catheter programme in 2007, before those patients were referred, mainly to Gothenburg which also receives referral patients from Umeå and Örebro. In Lund and Stockholm the basis for patient recruitment is by and large the regional catchment area. However, having in mind the surgical numbers of the two latter the surgical activity in Gothenburg and Linköping seems low. One could just speculate if there are differences in indications for surgical closure or if it is a result of a more successful catheter treatment. These are important quality issues of care needed to discuss and further analyse.

Quality of life before and after surgery and catheter intervention

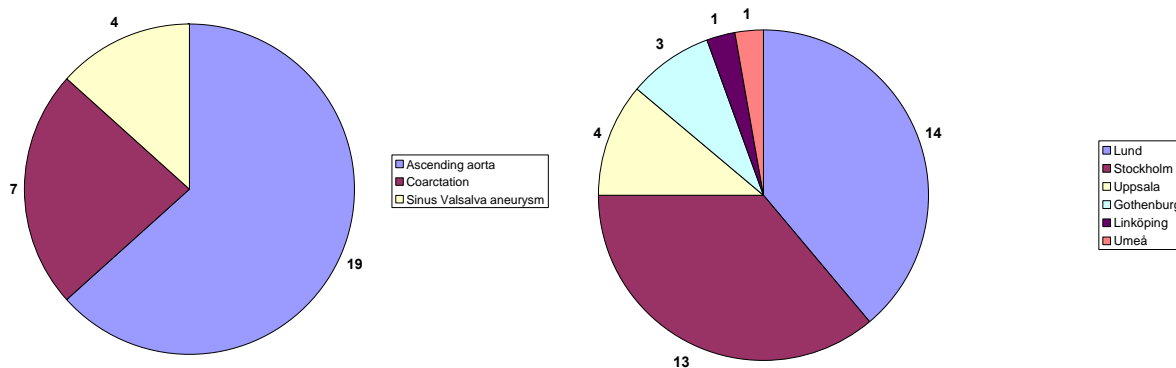
EQ-5D

		<u>Mean</u>	<u>Median</u>	<u>Range</u>
Catheter interventions (N=91)	Before	71	78	1-100
	After	79	80	20-100
Cardiac surgery (N=50)	Before	66	70	1-100
	After	65	70	3-100

In 141 patients who have had cardiac surgery or a catheter intervention during the period 2005-2007 there was data on quality of life in terms of EQ-5D both before and after the procedure. As a mean the quality of life assessment was made 128 days before and 240 days after the procedure. In the surgical group quality of life is unchanged while a slight improvement can be seen after the catheter intervention. However, it must be realised that the underlying condition, functional capacity etc. differ greatly between the groups making interpretation difficult.

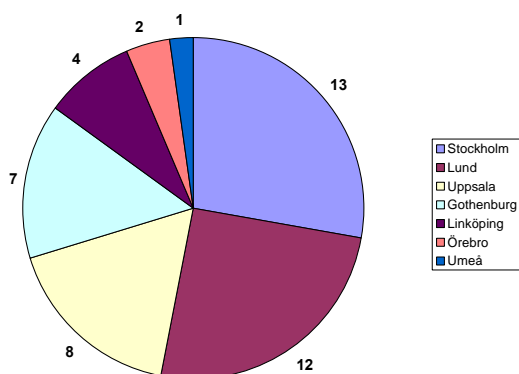
Cardiac surgical measures 2006 – 2007. Distribution of types and performing centres

Aortic surgery (n=30)



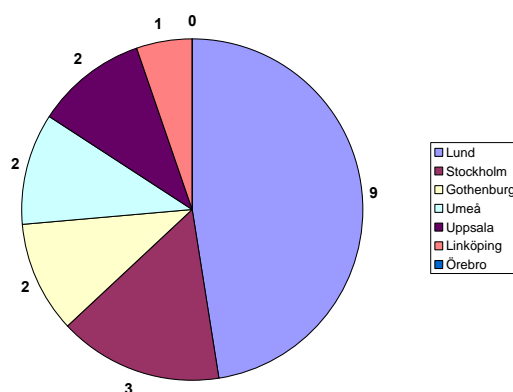
Surgery of the ascending aorta dominates aortic surgery. In aortic (re-)coarctation the number of catheter interventions (14 in 2007) outnumbers surgical intervention (7 during 2006-2007). Aortic surgery is performed in nearly all centres, however infrequent in most of them.

Aortic valve surgery, isolated or composite (n=47). Distribution of performing centres.



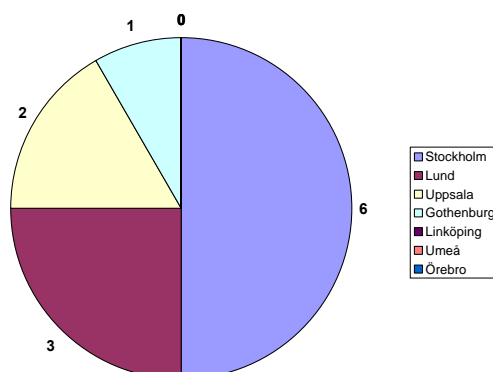
Aortic valve surgery, apart from closure of shunt lesions, is the most common type of surgery. Besides aortic valve replacements (mechanical and biological prostheses, homografts) it also includes Ross procedures and valvuloplasty.

Mitral valve surgery (n=19). Distribution of performing centres.



Mitral valve surgery may also include surgery on the systemic AV-valve in transpositions. In 8 cases valvuloplasty, in 3 cases anuloplasty and in 8 cases valve replacement have been made. Nearly half of these procedures were made at one centre.

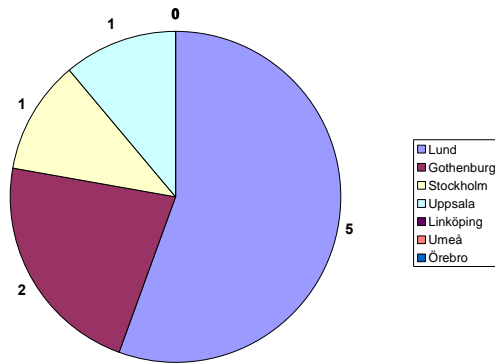
Tricuspid valve surgery including Ebstein's anomaly (n=12). Distribution of performing centres.



3/4 of tricuspid valve surgery have been performed in two centres. In 8 of the 12 procedures valve replacement took place. Valvulo- or anuloplasty was not performed at the two centres with the lowest number of procedures.

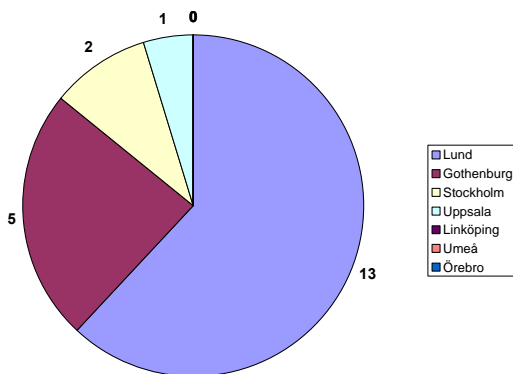
Surgery of complex lesions. Distribution of performing centres.

Subvalvular aortic (n=5) and pulmonary (n=4) obstruction

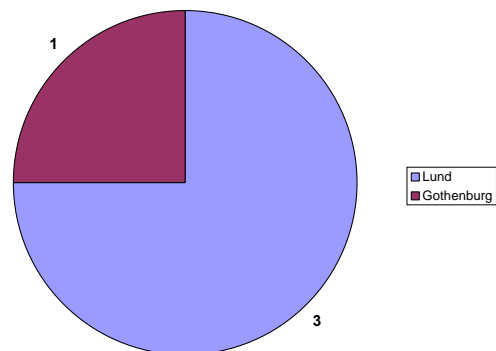


Surgery in complex lesions is rather infrequent. In spite of this and the complex nature some interventions are performed outside the two centres of paediatric heart surgery.

Conduit (n=16) and Fallot (n=5) surgery



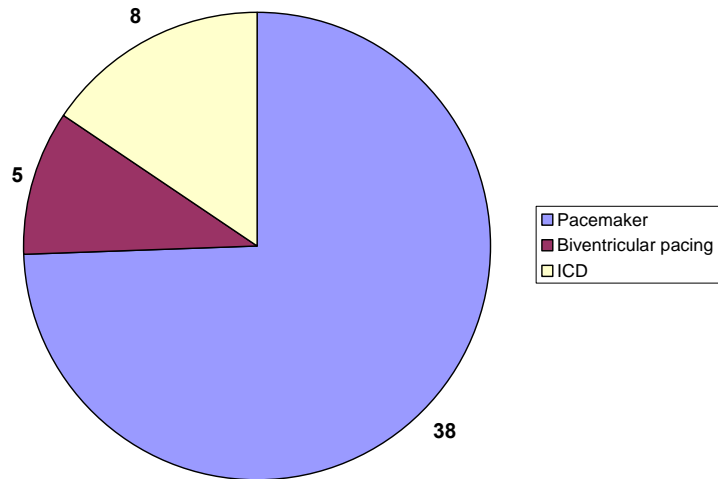
TCPC (n=4) surgery



In Sweden, cardiac surgery in grown-up congenital heart disease is not centralised like its paediatric counterpart. It seems that anatomically not straight “simple” lesions and even very complex ones are occasionally operated outside the two recognised centres of paediatric heart surgery. Only in extremely complex lesions like TCPC (total cavo pulmonary connection) surgery and conversion of Fontan states the patients are directed to the specialised surgical centres. Cardiac transplantation in adults with congenital heart disease (3 cases during 2006 – 2007) has so far taken place in Lund and Gothenburg which until recently were the only two transplanting centres in Sweden.

What impact this lack of centralisation has on surgical results is unknown and would deserve an investigation of its own as the registry does not cover surgical complications. However, this may be solved in future with the incorporation of paediatric cardiology and heart surgery in the registry.

Pacemaker and ICD



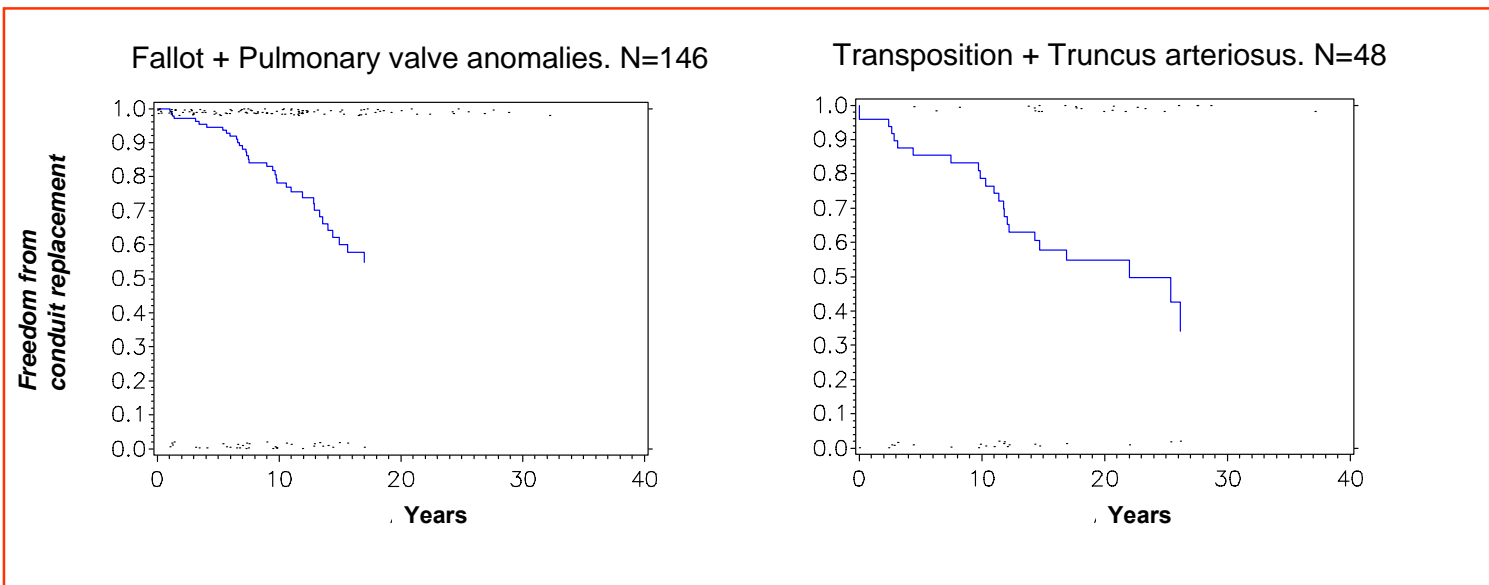
During 2006 – 2007 there were 51 pacemaker or internal cardiac defibrillator (ICD) implantations or renewals (of leads or battery). The number of ICD:s is small but increasing compared to former years. The role of biventricular pacing for heart failure in congenital heart disease is still unclear but, as seen, it has been applied in some patients.

CONDUIT

Conduit is a structure, usually including a valve, to connect nonadjacent parts of the cardiovascular system. In a majority of patients this is between the right ventricle and the pulmonary artery. In Sweden homografts have been more or less exclusively used for this purpose. In complex transpositions and truncus arteriosus the position of the conduit is “non-anatomic”. Although not a conduit in a proper sense, homografts are also used to overcome pulmonary valve disease, particularly when pulmonary regurgitation develops after surgical repair of tetralogy of Fallot or pulmonary valve stenosis. In those cases the homograft (sometimes just the valve is used) is in an anatomic position. With time the conduit degenerates and needs replacement. It has therefore been predicted that conduit replacement would be an important part of the future cardiac surgery in grown-up congenital heart disease. However, percutaneous catheter pulmonary valve insertion has developed and started in Sweden during 2007 (see catheter interventions - page 21). Analysis of conduit durability and survival is important; it is quality assessment of the treatment, it points out a standard level important to know when new therapies are introduced, it helps to predict future needs of interventions and it serves as a source of information to patients and caregivers.

In the registry there are 225 patients who have had conduit surgery, at any age. The numbers and the distribution of the underlying diagnoses are:

Fallot + pulmonary valve anomalies	146
Complex transposition + truncus arteriosus	48
Other	31



The graphs show the freedom from conduit replacement in the two main groups. Irrespective of conduit position, “anatomic” or “non-anatomic”, around 50 % of the patients have managed without conduit replacement the first 20 years after primary implantation. However, the incidence of early conduit replacement seems slightly more frequent in the transposition-truncus arteriosus group. Early replacement could suggest that surgical and technical factors, rather than degeneration, are more important when the conduit is in a “non-anatomic” position.

In an international perspective the long-term results of Swedish conduit surgery, as described here, maintain a very high standard.

PULMONARY HYPERTENSION - THE EISENMENGER SYNDROME

In primary and scleroderma-associated pulmonary hypertension specific vasodilating drug therapy has proven to have symptomatic and prognostic benefits. Hypertensive pulmonary vascular disease is not an uncommon complication of congenital heart disease. There has been a fear that vasodilators in this setting would cause worsened hypoxemia as they also act on the systemic circulation. However, this has turned out not to be true and there are even reports suggesting that endothelin-antagonism reduces pulmonary vascular resistance and improve working capacity (6 minutes walk test) in this subset of pulmonary hypertension. As these therapies are expensive it seems rational to describe the patient population and analyse to what extent specific drug therapy already have been initiated before general decisions and recommendations are made

In the registry "secondary pulmonary hypertension" has a diagnostic code of its own. At the end of 2007 there were 161 patients with this diagnosis, eg 2,6% of registered patients.

	N	Alive	Patients with Down's syndrome
All	161	117	33
<u>Underlying diagnosis</u>			
Shuntlesion	112	80	31
of which:			
ASD	33	26	1
VSD	69	47	30
Other shunt	10	7	0
Transposition	9	3	0
Fallot/RV anomali	5	5	0
Other	35	29	2

More than 2/3 of the patients have an Eisenmenger syndrome, a shunt lesion complicated by hypertensive pulmonary vascular disease with bidirectional/reversed shunt. Ventricular septal defect (VSD) is the most common type of underlying shunt lesion, followed by atrial septal defect (ASD). In the VSD group nearly half of the patients have Down's syndrome. Mortality is high, 27 % of the patients have died. More than 2/3 of the deaths were cardiac. The median age at death was 37 (range 20-79) years. According to an earlier analysis mortality rates in Down's syndrome in this context did not differ from non-Down.

Specific drug therapy for pulmonary hypertension (Ca-antagonism excluded)

During 2007 there are data on drug therapy for only 48 of the 117 patients with pulmonary hypertension who are alive. The reason for absent data may be that the patient has not been seen during 2007 or is cared for at a lower level than the specialised unit. It is an impression that the latter often is the case of patients with Down's syndrome.

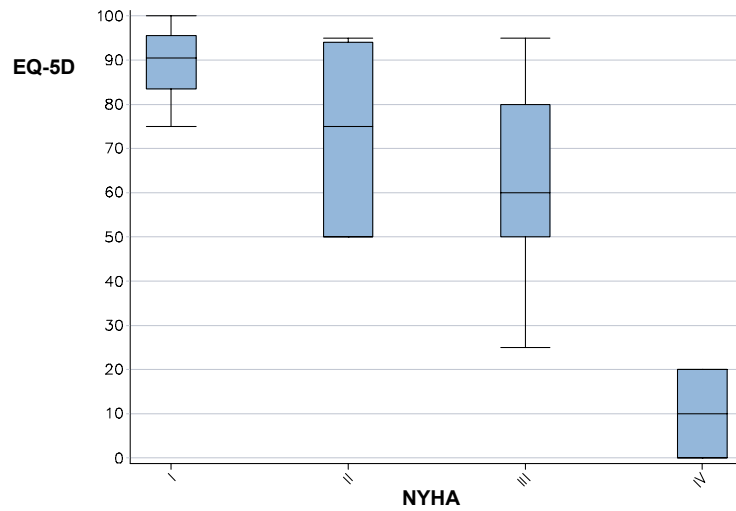
	N	Endothelin-antagonist	Sildenafil	Prostacyclin
ASD	12	7	1	1
VSD	15	1	1	0
Other shunt	4	1	0	0
Other lesion	17	4	2	1

NYHA classification in those alive (n=117) at the end of 2007 related to the presence of Down's syndrome and the use of specific drug therapy related to functional class

<u>NYHA</u>	<u>Down's</u>	<u>Non-Down's</u>	<u>Patients with specific therapy for pulmonary hypertension</u>
I	2	20	1
II	2	21	5
III	11	30	11
IV	1	1	0
Not able to classify/Unknown	8	21	1

In general the treatment intensity is low. The highest treatment frequency is found in the ASD group and the reason for this is not clear. The prevalence of specific therapy for pulmonary hypertension is rather equal in NYHA II and III, while it is lower in those less symptomatic. However, the NYHA classification refers to the latest appointment and not necessarily the state before treatment. Compared to 2006 the number of treated patients has increased, then only 3 ASD patients and no VSD patient received endothelin-antagonists.

Quality of life – EQ-5D - related to NYHA functional class at the latest appointment



In 55 patients with the diagnosis of “secondary pulmonary hypertension” there were data on EQ-5D rating as well as the NYHA functional class. As in general, the quality of life is associated to the functional capacity. Furthermore, it could suggest that improvement of functional capacity, by means of specific drugs, would lead to a higher quality of life.

QUALITY ASSESSMENT OF GIVEN CARE

Below are some examples of routines that would be expected in a service with a high standard of care. The registry provides information on to what degree these demands are fulfilled. When not satisfactory feed-back is given and action for improvement implemented.

Renal function assessment in ACE-inhibitor/AII-antagonism treatment

713 registered patients are treated with either an ACE-inhibitor or an AII-antagonist. Because of the pharmacological action of these drugs the renal function should regularly be checked. In 549 (77%) there was no information about renal function (plasma-creatinin) at the latest visit or the visit before that. It could indicate suboptimal care and should be discussed in the working group.

Haematological assessment in cyanosis

Regular determination of the haemoglobin concentration should be a part of the clinical evaluation in patients with cyanotic heart disease. Arterial oxygen saturation was reported below 90 % in 118 patients at their latest visit. In only 58 of them (49%) there was a coexisting report of the haemoglobin concentration. The present routines seem suboptimal and have to be discussed and improved.

Blood pressure measurement in arm and leg in patients with repaired aortic coarctation

As shown in the section on coarctation (see above) blood pressure measurements in both arm and leg are lacking in around 30 % of patients with repaired aortic coarctation. However, due to the feed-back of registry data this is an improvement compared to 2005. Arbitrary, > 90% of the patients should have the blood pressure in the upper and lower part of the body estimated and there is still a potential for further improvements.

QRS-duration in tetralogy of Fallot

In tetralogy of Fallot a QRS-duration > 180 ms in the ECG is a risk marker for serious ventricular arrhythmia and sudden death. A high standard of care in tetralogy of Fallot would therefore include assessment of the QRS-duration. Since 2003 this has been the case in 85-96% of the patients, with the highest figure in 2007. This is a very satisfactory level.

The “One stop shop” concept, development of performance rates 2005 - 2007

The “one stop shop” performance means that all necessary investigations are made and the results available at the same day as the doctor’s appointment. There are several advantages of this. The patient receives an immediate and comprehensive evaluation and doctor’s conclusions thereby reducing speculative negative psychological mechanisms. Many of the patients have a professional life and, spending one instead of several days for investigations reduces the time away from work.

In the analyses below, covering 2005 - 2007 patients that had just one doctor’s visit during the given year were chosen, this because it would have been very difficult from an analytical point of view to include those with multiple yearly appointments. Then it was to decide when an examination besides the doctor’s visit was an essential part of the clinical assessment; this was defined as an investigation – ECG, echocardiography, exercise test or chest X-ray – taking place within 90 days before or after the doctor’s visit. The next step was to analyse if these essential investigations had been performed on the same day as the doctor’s visit or not.

National data

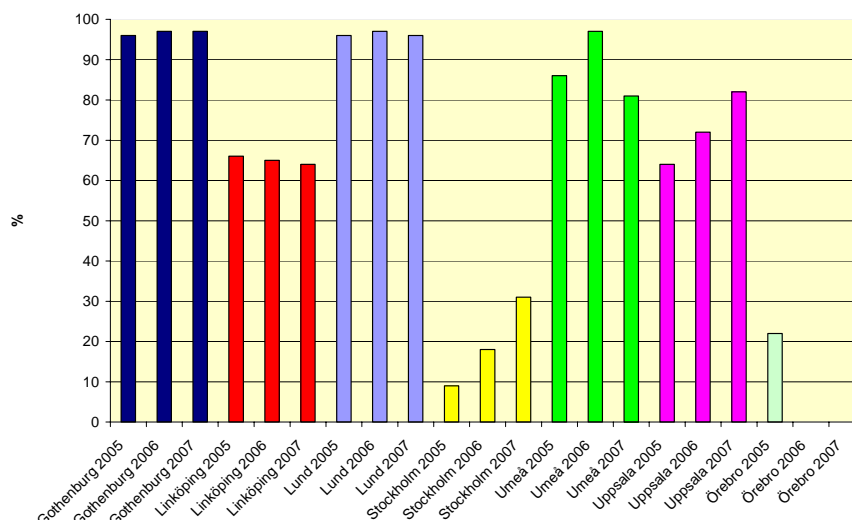
	<u>2005</u>	<u>2006</u>	<u>2007</u>
Patients with just one doctor's visit	1486	1902	1920
Patients who only had ECG	253	300	306
% of those who had it the same day as doctor's visit	96	94	97
Patients who had both ECG and Echocardiography	691	956	990
% of those who had both the same day as doctor's visit	81	78	77
Patients who had 1 exam*, ECG excluded	855	1143	1143
% of those who had it the same day as doctor's visit	75	73	76
Patients who had 2 exams*, ECG excluded	256	314	316
% of those who had both the same day as doctor's visit	34	22	29
Patients who had 3 exams*, ECG excluded	12	7	5
% of those who had all 3 the same day as doctor's visit	0	0	0

*=Echocardiography, exercise test or chest X-ray

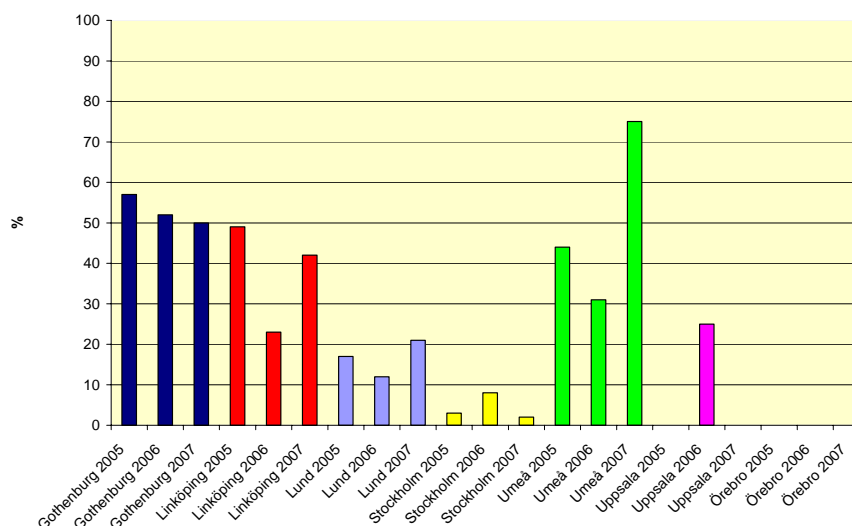
ECG is recorded nearly always in conjunction with the doctor’s visit. In those where echocardiography together with an ECG were thought essential $\frac{3}{4}$ had the two exams on the same day as the doctor’s appointment. Since 2005 the figures have declined rather than improved. With increasing number of essential exams the ability to have them on the same day as the doctor’s visit diminishes rapidly. In the low number of patients who needed an echocardiogram, an exercise test and a chest X-ray it seemed impossible to concentrate all three on the same day as the patient met the physician. However, one must recognise that sometimes the need for an investigation is dictated by new or changing symptoms revealed at the doctor’s visit. Thus, it is difficult to determine what frequency of “one stop shop” that defines optimal care and good logistic organisation. One way would be to use the single centre with the highest frequency as a standard.

“One stop shop” performance rate 2005 – 2007 per centres

Patients with one examination besides ECG



Patients with two examinations besides ECG



When just one investigation besides ECG is necessary it is often echocardiography. Three centres manage in around 90 % or more of the patients to apply the “one stop shop” concept. A common feature for these centres is that the cardiologists perform the echocardiogram themselves. In Stockholm and Uppsala the proportion of “one stop shop” visits has increased during the last years. It is probable that echocardiography in the hands of the responsible cardiologist seems important from the point of “one stop shop”.

In patients with two investigations besides ECG, Umeå performed best during 2007. Interestingly, the Umeå region covers a very large area which makes long-distance travel common and it is not unlikely that this has stimulated better logistics. Next to Umeå, Gothenburg with an openly declared ambition to apply the “one stop shop” concept has the highest rates in patients with two essential investigations. These two examples seems to demonstrate that well thought-out logistics make it possible to, make care more efficient and less time-consuming for the patients.