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Mr. Speice

Independent Study and Mentorship- 3A

14 October 2016

Research Assessment 4

Subject: Pediatric Surgery

MLA Citation:

Works Cited

McMillan, Julia A. "Pediatric Surgeons: Partners in Care for Children." *Health and Wellness Resource Center [Gale]*. Mackinvia, Jan. 2012. Web. 14 Oct. 2016.

"Pediatric Surgery." *Health and Wellness Resource Center [Gale]*. Mackinvia, 2013. Web. 14 Oct. 2016.

Nieminen, Heta, Heikki Sairanen, Tero Tikanoja, Markku Leskinen, Henrik Ekblad, Paivi Galambosi, and Eero Jokinen. "Long-term Results of Pediatric Cardiac Surgery in Finland: Education, Employment, Marital Status, and Parenthood." *Health and Wellness Resource Center [Gale]*. Mackinvia, Dec. 2003. Web. 14 Oct. 2016.

Assessment:

When deciding my subject for this research assessment I knew I was curious to learn more about general pediatric surgery and also the effects of pediatric surgery on a patient's life. With those interests in mind I found two articles on pediatric surgery and one on a study of the long term results pediatric cardiac surgery.

The first article was quite different than what I expected of an article about pediatric surgery. I expected it to just be general facts of pediatric surgery; however, it spoke of the lack of

pediatric care in rural areas and the meaning of being a pediatric cardiac surgeon. This information spoke to me on the impact I could make as a pediatric surgeon. The lack of pediatric surgeons in rural areas shows that there is availability within this field which makes me feel more confident in wanting to go into this field. This also shows how this career would impact a rural area, something I hope to do with this career. The article also speaks of the challenges that come with pediatric surgery which is also something that interests me about a future career. This article add to my previous knowledge I had before, something new that I found very interesting was the mission statement of the American Pediatric Surgical Association which depicts what being a pediatric surgeon is all about. The second article is mostly facts on pediatric surgery. I found that most of this article had irrelevant information to my topic of study, since it spoke on specific conditions of other pediatric surgery subspecialties. Overall both of these articles added to my overall understanding of pediatric surgery. I am now able to clearly see that this is a career I would be successful in and that truly does interest me, but also helped me evaluate that I do want to keep learning about the specialty of pediatric cardiac surgery.

The third article spoke about the effects of pediatric cardiac surgery beyond the patient's simple survival. This is relevant to me because I am searching for a career that will help me impact people's live and I still had not truly learned about the aspects of pediatric surgery beyond the simple survival of a patient. I was able to see that most patients have a relatively normal life which is one of the most rewarding parts of this career. I have been discouraged in the past to want to become a pediatric cardiac surgeon due to the difficulty and stresses of the job, but I believe the rewarding parts of the career, like saving a patient's life and seeing him/her live a normal and healthy life, makes it all worthwhile. Reading a study which mostly shows how pediatric cardiac surgery leaves a positive impact on a patient's life adds to my want to study this

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career. All the information in this article will help me continue to be inspired to follow this career and to understand the patient's side of having pediatric cardiac surgery. Both of these articles have brought me to questions the patient side of pediatric surgery, and to learn more about the lack of pediatric care in rural areas.

**Pediatric surgeons: partners in care for children.**

Julia A. McMillan. *Contemporary Pediatrics*. Jan 2012 v29 i1 p7(1).

**Comment [1]:** I wanted to research about just the basics of pediatric surgery, since pediatric cardiac surgery is a specialty.

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According to the American College of Surgeons Health Policy Research Institute, in 2006 only 12.8% of counties in the United States had a single **pediatric surgeon** of any type (general or subspecialty), and only a little more than half of those counties had a **pediatric general surgeon**. (1)

Children in rural counties are particularly poorly served by **pediatric** surgeons. In 2006, 97% of rural counties lacked a **pediatric surgeon**. For those of us fortunate enough to work with well-trained **pediatric** general surgeons and **pediatric** surgical subspecialists on a daily basis, it's difficult to imagine caring for infants and children whose congenital, traumatic, infectious, or oncologic conditions require surgical intervention without a **surgeon** trained and inclined to provide care for **pediatric** patients and to support their families.

The article in this issue by Jeffrey Lukish, MD, on minimal access surgery, as well as the article by Thomas Inge, MD, and Linda Kollar, RN, on bariatric surgery for adolescents in the October issue, highlight the importance of distinct considerations when infants and children require or are being considered for surgical interventions.

As Inge and Kollar emphasized, collaboration among pediatricians, **pediatric** subspecialists, and **pediatric** surgeons is essential when evaluating an adolescent for possible bariatric surgery.

Lukish describes concerns for safety and the particular need for right-sized surgical instruments in extrapolating what has been learned about minimal access surgery in adults to infants and children.

Adoption of these techniques and procedures by **pediatric** surgeons requires care and patience as well as thoughtful consideration of their risks and benefits.

Dissemination of experiences with new approaches through publications that reach colleagues throughout the country and the world is essential, so that benefits as well as unanticipated pitfalls can be recognized.

I'm afraid that pediatricians sometimes think we are the most vocal and important advocates for improved health and access to medical care for children. It's true that the approximately 60,000 pediatricians present a larger voice for advocacy than the approximately 3,000 **pediatric** general surgeons and **pediatric** surgical subspecialists. However, our **pediatric** surgical colleagues are

**Comment [2]:** The lack of pediatric surgeons means there is availability for students who hope to become pediatric cardiac surgeons.

**Comment [3]:** Shows how this is a career that will leave an impact in the world, the increase of pediatric cardiac surgeons will save many lives.

**Comment [4]:** The difficulty in any pediatric surgery specialty is something that is very apparent and spoken of; however this challenge is what I find most interesting in any pediatric surgery specialty.

important teammates in our efforts to enhance recognition of the distinct needs of infants and children.

The mission statement of the American **Pediatric** Surgical Association, "To Ensure Optimal **Pediatric** Surgical Care of Patients and Their Families, to Promote Excellence in the Field, and to Foster a Vibrant and Viable Community of **Pediatric** Surgeons," highlights the importance of **pediatric** surgeons as partners in making sure that our patients receive the most expert care possible.

#### REFERENCES

(1.) Poley S, Ricketts T, Belsky D, Gaul K. **Pediatric** surgeons: subspecialists increase faster than generalists. Bull Am Coll Surg. 2010;95(10):35-38.

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Contemporary Pediatrics

**Record Number:** A456582027

**Comment [5]:** This mission statement is something I have never seen before, but depicts what I will be doing.

## Long-term results of pediatric cardiac surgery in Finland: education, employment, marital status, and parenthood.

Heta Nieminen, Heikki Sairanen, Tero Tikanoja, Markku Leskinen, Henrik Ekblad, Paivi Galambosi, Eero Jokinen. *Pediatrics*. Dec 2003 v112 i6 p1345(6).

### Author's Abstract:

**Objective.** This population-based study was designed to examine the psychosocial outcome of Finnish patients who had been operated on for congenital heart disease during childhood.  
**Methods.** A questionnaire was mailed to 3789 adult patients who had been operated on for congenital heart defects in Finland. Of these, 2896 (76%) answered the questionnaire. The mean age of patients was 33 years (range: 18-59 years), and they had had their first operation 9 to 46 years earlier.

**Results.** The patients had coped well with their defects when compared with the general Finnish population. The educational level of patients was comparable to and employment level was higher than expected (70% vs 66%, respectively). They were living in a steady relationship as often as the general population, but the number of parents among the patients was lower than that expected (47% vs 49%, respectively). The incidence of congenital heart disease among the 2697 children of the patients was 2.4%.

**Conclusion.** Our results confirm that in addition to high survival rate, the long-term psychosocial outcome of patients with surgically treated congenital heart defects is good if they do not have any additional syndromes that cause mental retardation. *Pediatrics* 2003;112:1345-1350; heart defects, congenital, population, education, employment, marital status.

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The progress of medicine has allowed an increasing number of congenital heart defects to be corrected or alleviated surgically while maintaining high overall survival rate. In fact, most patients, especially those with simple defects, have a normal life expectancy after a successful operation. (1) However, from the patient's point of view, survival may not be the most important measure of therapeutic success. A long life burdened with disability and dissatisfaction can be of

**Comment [6]:** I picked this article because I want to learn about the impacts a pediatric cardiac surgeon has on a community.

**Comment [7]:** This paragraph shows a pediatric surgery can not only save someone's life but can bring the patient a long and normal life.

questionable value to a patient who, by survival criteria, represents a good outcome. To evaluate our operative results beyond survival, we examined the basic psychosocial aspects of life, such as education, employment, and marital status, by sending a questionnaire to all adult patients who had been operated on for congenital heart defects in Finland. The patient data came from the Finnish national research registry of **pediatric cardiac surgery**. (1) The results were compared with the age-matched general population.

#### METHODS

During the years 1953 to 1989, a total of 7240 **cardiac** operations were performed on 6461 children in Finland. A total of 3842 patients had reached the age of 18, and 3789 of them had a known address on October 28, 1998. All of these adult patients were included in this study. In addition, 226 patients who were operated on in Finland may have reached adulthood, but their outcome was not known as they were either not identified from population registry (84) or had emigrated (142).

Each operating hospital sent the inquiry to its own patients, and the results were combined. All units used the same questionnaire designed to provide information about the patient's general state of health, exercise tolerance, education, occupation, marital status, offspring, and **cardiac** follow-up. The questionnaire was designed to be easy and quick to complete and unambiguous to interpret. The patients were asked to classify their general state of health as good, moderately good, or poor and to list their concurrent illnesses. The patients estimated their exercise tolerance by classifying their physical fitness as normal, less than normal, or significantly less than normal and by describing their ability to climb stairs and run. The investigators translated the patients' answers into the New York Heart Association (NYHA) classification. Because the questions did not differentiate between NYHA classes I and II, these classes were combined for the analysis.

We classified education into 3 levels: compulsory, vocational, and university. Compulsory education in Finland consists for 9 years of comprehensive school. The vocational level includes graduation from vocational school or high school. The university level required a degree from an institute of postsecondary education.

The patients were divided into different groups by the main diagnosis according to a previously published hierarchy. (1) The 9 most common diagnoses were patent ductus arteriosus (PDA), coarctation of the aorta (CoA), atrial septal defect (ASD), ventricular septal defect, tetralogy of Fallot (TOF), transposition of the great arteries (TGA), pulmonary stenosis (PS), valvular aortic stenosis (AS), and univentricular heart (UVH). The patients with TOF, TGA, and UVH were combined into 1 group (cyanotic) to evaluate the impact of a complex defect on coping.

**Comment [8]:** Although this seems pretty obvious and understandable I had not really thought of this burden as a reason people did not want to have cardiac surgery.

**Comment [9]:** I want to learn the effect of pediatric cardiac surgery in a persons life beyond survival

### Statistical Analysis

The educational, employment, and family status were compared with that of the general population, using data from the national statistical center, Statistics Finland. The expected values were calculated as weighted averages of published age- and sex-specific rates. The calculations were made separately for each patient group. In the education section, the patients who were younger than 20 years were excluded. Unpaired t test was used to compare the ages of male and female patients; the  $\chi^2$  test was used in comparing of frequencies between patients and the general population.

### RESULTS

A total of 2896 patients (76%) completed and returned the questionnaire. Women answered the inquiry more often than men (80% vs 72%, respectively;  $P < .001$ ). The number of replies received from patients in each diagnostic group varied from 72% to 82% (Fig 1). The age and defect type distributions were similar between the patients who completed the questionnaire and those who did not. Henceforth, in this report, the term "patient" refers to patients who provided data.

The median age of the patients at first operation was 6.7 years (range: 0-15). Their median follow-up time was 25.6 years (range: 9-46), and their median age was 31.7 years (range: 18-59) at the time of data collection. Men had more complex defects than women: 67% of CoA, 56% of TGA, and 62% of AS patients were male. There was a female predominance among the patients with PDA (76%) and ASD (61%). Men were younger than women ( $P < .001$ ) with a median age of 30.8 years (range: 18-57); the median age of women was 32.5 years (range: 18-59). Of the 3789 patients, 100 (2.6%) had received a diagnosis of mental retardation. Seventy-six questionnaires were received from them, in most cases completed by their guardians.

### General State of Health

Most patients (2227 [77%] of 2896) described their general health to be good. One fifth of the patients (611 [22%]) classified their health as moderately good, and only 36 patients (1%) believed their condition to be poor. In most cases, poor health was attributable to a condition unrelated to the **cardiac** defect or its treatment. Surprising was that no patients with UVH ( $n = 22$ ) or TGA ( $n = 64$ ) reported poor health. Five patients with PDA ( $n = 1066$ ) could be considered to be in poor condition because of their heart defect.

### Exercise Tolerance

Most patients assessed their exercise tolerance to be good. A vast majority, 97% (2320 of 2896), were classified into NYHA I and II; 68 patients (2%) were in NYHA III. Only 7 patients were symptomatic at rest (NYHA IV). The distribution was similar between diagnostic groups,

**Comment [10]:** Another positive effect of pediatric cardiac surgery



except for the TOF (n = 180) and UVH groups. Fifteen patients (8%) had TOF, and 5 patients (23%) had UVH in NYHA class III; 1 patient from the TOF group and 1 from the UVH group were classified as NYHA IV.

#### Education

The educational profile of the patients was comparable to the expected (Table 1). Most of the patients (2100 [79%] of 2676) had more than compulsory education, and 10% (257 of 2676) had a university degree. The level of education was significantly different from the general population in 3 patient groups. The number of patients with a vocational or university degree in the CoA group (356 [85%] of 420) was 6% higher than expected (P = .003). In the ASD group, more patients than expected had compulsory education only (24% [101 of 421] vs 21%, respectively; P = .038). The educational level was low also among the patients with a cyanotic defect. Of the 235 patients, 63 (27%) had compulsory education only, as compared with the expected 21% (P = .025). When the 13 patients with mental retardation (12 with TOF, 1 with TGA) were excluded, the proportion of patients with only compulsory education decreased to 22%, which was comparable to the expected frequency. A university degree was less common than expected (3% vs 7%; P = .014) among patients with cyanotic defects. The exclusion of patients with mental retardation did not eliminate this difference.

#### Employment

The employment rate (70%) was higher among the patients than in the general population (66%; P .001; Table 2). The patients with PDA and CoA (n = 456) were employed significantly more often than their reference populations (77% vs 69% and 74% vs 66%, respectively; P < .001). As seen in Table 2, the patients with cyanotic defects were employed less often than expected. The difference disappeared when the patients with mental retardation were excluded. The overall unemployment rate of only 6% among the patients contrasted with the expected rate, which was nearly twice as high (11%; P < .001). The proportion of students was similar to the general population. The observed retirement rate was 7% and the expected was 3%. Only 38 of the 207 retired patients were clearly disabled because of their heart defect. The most common reason for retirement was mental retardation, in 66 patients.

#### Marital Status

The number of married patients was comparable to the general population. A total of 1043 (36%) patients were married, women more often than men (40% vs 30%, respectively; P < .001; Table 3). The proportion of patients who were unmarried but living with a partner was greater than expected (23% vs 20%, respectively; P < .001). However, there also were more single patients than expected (37% vs 35%; P = .048). The concurrency of a high number of

patients living in a relationship and a high number of singles was possible because the divorce rate among the patients was lower (4%; 105 patients) than the expected 7%. There was not significant difference in the marital status between the cyanotic group and the general population, except the low divorce rate of patients (1% vs 5%).

#### Offspring

The patients had become parents less often than expected. The difference was similar for both sexes (Table 4). The number of children in a family was comparable to that of the general population, except that there were only 45 female patients who had 4 or more children. The difference to the expected 67 was statistically significant ( $P = .006$ ). The number of parents with PDA (620 of 1066) and PS (60 of 123) was lower than expected (58% vs 61% [ $P = .032$ ] and 49% vs 58% [ $P = .045$ ], respectively). The female patients with cyanotic defect became mothers less often than expected (Table 4). There were only 45 mothers (35%), while the expected proportion was 46% ( $P = .008$ ). The difference diminished ( $P = .039$ ) but did not vanish when the patients with mental retardation were excluded.

The incidence of congenital heart disease (CHD) among the offspring of the patients was 2.4 times that of the general incidence of approximately 1%. (2) Of the 2697 children, 64 (2.4%) had a heart disease. Women had a higher risk of having children with CHD (Table 4). The incidence of CHD in the offspring was 2.7% for women and 1.6% for men ( $P < .001$ ). Relatively high risk of recurrence of CHD was seen in the PDA (2.7%), ASD (2.6%), and PS (5.8%) groups ( $P < .001$  in all). Two patients with PDA had 3 children and 3 mothers (2 PDA, 1 PS) had 2 children with CHD.

**Cardiac** Follow-up

**Comment [11]:** possibly because CHD can be hereditary

Of the adult patients, only 26% (742) were in **cardiac** follow-up. After exclusion of the patients with PDA and ASD there were still 53% (724 of 1374) of patients without follow-up. Most patients (78%; 564 of 724) who had dropped out of follow-up believed themselves to be in good condition and in NYHA class I to II. The only patient group in which all patients had regular appointments was UVH. Several patients with known high residual risk were not seen regularly; for instance, 36% of TOF and 60% of CoA patients and even 2 patients after Mustard operation for TGA.

#### DISCUSSION

This population-based study reveals the late results of **pediatric cardiac surgery** beyond survival curves. A comprehensive and accurate national research registry of **pediatric cardiac surgery** in Finland (1) guarantees the reliability of the results. With good cooperation between hospitals, we were able to include 3789 patients in the study and receive a completed questionnaire of the basic elements of psychosocial life from 2896 adult patients. The national population registry and statistics center gave us the opportunity to compare patients with the general population, which is recognized as the most relevant control group (3). To maintain a high completion rate, we designed the questionnaire to be simple. The decision was made to address only the basic elements of life: health, fitness, education, employment, and family. Hence, we were able to achieve a relatively high reply rate of 76%, yet the most important and unavoidable bias of this study is the inability to define the difference between the patients who answered and who did not. Another important bias is that the results are based solely on the patients' own report of their conditions. This might have given slightly overestimated values toward the awaited "good results." For example, if patients had been retired and a student at the same time, then it is more likely that the answer to the employment question would be student rather than retired.

A chronic disease has been reported to produce both negative and positive effects on the patient's life. (3,4) In many studies, good coping has resulted in an excellent quality of life, even for severely physically disabled patients. (5-7) The documented positive effect usually relates to education and employment, (5-9) whereas difficulties in reaching independence and finding a partner are the most common negative effects reported. (8,10)

Subjectively, the vast majority of the patients estimated their health in general to be good. Poor health was usually associated with noncardiac conditions. The overall health of patients within different diagnosis groups has previously been reported to be equal or better than in our material. (11-14)

The overall exercise tolerance of the patients who were operated on also was good. Most patients

were assigned to NYHA classes I and II in all diagnostic groups. Our results were comparable to previous reports in patients with CoA, ventricular septal defect, PS, AS, and TGA. (11,15-17) Both similar (18,19) and better (20,21) exercise tolerance has been reported previously after TOF repair. In our results, the patients with late corrective **surgery** or only palliative **surgery** worsened the estimated exercise capacity.

The patients had coped surprisingly well with their defects. The level of education was equivalent, the employment level was higher than expected, and they were living in a steady relationship as often as the general population.

A high level of education has been reported frequently in several studies of different diagnostic groups. (7,11,17,22) After exclusion of the patients with mental retardation, the number of patients with vocational or university education was higher than expected. However, we found that patients with cyanotic defects were less likely to have graduated from a university. Whether this was caused by lack of encouragement or a decreased capacity for performance remains open.

The patients clearly were employed more often than expected. Even the exclusion of the most frequently employed group, the PDA patients, did not change this positive result. The patients with cyanotic defects were not employed as often as expected, but after exclusion of patients with mental retardation, the number was comparable to the general population. The low unemployment rate reflects the high value of work among these patients, a finding that has been reported previously. (8,9)

Marital status of the patients was comparable to normal. We do not know how many of the single patients were still living with their parents and thus could not entirely contradict the previous idea of a low level of independence. (8,23) The low divorce rate could be a sign of either a successful relationship or overadherence and lack of independence. Our data do not provide an answer to this interesting matter.

The patients did not have children as often as the general population, a tendency seen also among the women in the Second Natural History Study. (11) It was expected that the women with cyanotic defects would often be childless, but the large number of patients (both female and male) without children in the PDA group was surprising.

The risk of having a child with congenital heart defect was 2.4 times higher than expected. The recurrence of a CHD varies widely between different defects. The patients with TOF and TGA have repeatedly reported not to have children with CHD, (20,24-27) whereas the patients with minor lesions have had children with CHD approximately 3 times (17,26,28) more often than expected. The overall risk of only 2 to 3 of 100 children can be considered small enough to encourage these patients to have offspring. However, they should also be provided with sufficient genetic counseling.

When patients with mental retardation were excluded, the proportion of those retired and having only compulsory education dropped to the level present in the general population. The effect of mental retardation in quality-of-life studies has to be considered carefully, because the retardation is usually not a consequence of the heart disease but can have an overwhelming effect on the quality of life. Furthermore, a sufficient degree of mental retardation precludes self-assessment of the quality of life, in which case its evaluation becomes dependent on a third party.

The frequent lack of adequate **cardiac** follow-up surfaced as a serious problem among this patient group. In this respect, our results are comparable to previous studies. (11,13,20) The reason for the high rate of follow-up dropouts likely is attributable to unsuccessful transfer of care from **pediatric** to adult clinic. The potential for hazardous consequences from lack of follow-

up has been documented previously. (29)

CONCLUSIONS

In the Finnish culture, it is appreciated, for both sexes, to have more than compulsory education, to be employed, and to start a family. When evaluated with these goals in mind, the results of **pediatric cardiac surgery** are very good. The good results should encourage us to treat the patients with congenital heart defects intensively even when the treatment is costly and often long-lasting.

Approximately half of the patients who were operated on for congenital heart defect should be **in cardiac** follow-up for the rest of their lives. However, in our material, 53% of patients who needed follow-up had dropped out. There should be an intense effort to develop an efficient chain of services to ensure the transition of patients and expertise from **pediatric** to adult **cardiac** follow-up.

ABBREVIATIONS. NYHA, New York Heart Association; PDA, patent ductus arteriosus; CoA, coarctation of the aorta; ASD, atrial septal defect; TOF, tetralogy of Fallot; TGA, transposition of the great arteries; PS, pulmonary stenosis; AS, valvular aortic stenosis; UVH, univentricular heart; CHD, congenital heart disease.

TABLE 1. Education Level of Patients Compared With Expected Level

All (n = 2676)				
	Patients		95% CI	Expected
Compulsory	576	21%	20-23%	21%
Vocational	1843	69%	67-71%	70%
University	257	10%	8-11%	9%
Cyanotic * (n = 235)				
	Patients		95% CI	Expected
Compulsory	63	27% ([dagger])	21-32%	21%
Vocational	164	70%	64-76%	72%
University	8	3% ([dagger])	1-6%	7%
Men (n = 1085)				
	Patients		95% CI	Expected
Compulsory	246	23%	20-25%	23%
Vocational	755	69%	67-72%	70%
University	84	8%	6-9%	7%
Men (n = 120)				

	Patients	95% CI	Expected
Compulsory	28	23%	16-31% 22%
Vocational	87	73%	65-80% 72%
University	5	4%	1-8% 6%

Women (n = 1591)

	Patients	95% CI	Expected
Compulsory	330	21%	19-23% 21%
Vocational	1088	68%	66-71% 70%
University	173	11% ([dagger])	9-12% 9%

Women (n = 115)

	Patients	95% CI	Expected
Compulsory	35	30% ([double dagger])	22-39% 19%
Vocational	77	67%	58-76% 72%
University	3	3% ([dagger])	0-6% 9%

CI indicates confidence interval

\* Patients with TOF, TGA, or UVH.

([dagger]) P < .05.

([double dagger]) < .01.

TABLE 2. Occupational Status of Patients Compared With Expected Levels

All (n = 2896)

	Patients	95% CI	Expected
Employed	2036	70% [parallel]	69-72% 66%
Unemployed	153	11% [parallel]	4-6% 11%
Students	375	13% [parallel]	12-14% 13%
Retired	207	7% [parallel]	6-8% 3%
Other *	114	4% [parallel]	3-5% 7%

Cyanotict (n = 266)

	Patients	95% CI	Expected
Employed	146	55% ([double dagger])	49-61% 61%
Unemployed	16	6% ([double dagger])	3-9% 10%
Students	50	19%	14-23% 18%

Retired	43	16% [parallel]	12-21%	2%
Other *	11	4% ([double dagger])	2-7%	9%

Men (n = 1188)

	Patients		95% CI	Expected
Employed	810	68%	66-71%	67%
Unemployed	75	7% [parallel]	5-8%	12%
Students	171	15%	12-16%	13%
Retired	107	9% [parallel]	7-11%	3%
Other *	16	1% [parallel]	1-2%	5%

Men (n = 136)

	Patients		95% CI	Expected
Employed	81	60%	51-68%	64%
Unemployed	11	8%	4-13%	11%
Students	24	18%	11-24%	2%
Retired	19	14% [parallel]	8-20%	2%
Other *	0	0% [section]		6%

Women (n = 1708)

	Patients		95% CI	Expected
Employed	1222	71% [parallel]	69-74%	65%
Unemployed	78	5% [parallel]	4-6%	11%
Students	204	12% ([double dagger])	10-13%	14%
Retired	100	6% [parallel]	5-7%	2%
Other *	98	6% [parallel]	5-7%	8%

Women (n = 130)

	Patients		95% CI	Expected
Employed	64	49% ([double dagger])	41-58%	59%
Unemployed	5	4% ([double dagger])	1-7%	9%
Students	26	20%	13-27%	19%
Retired	24	19% [parallel]	12-25%	2%
Other *	11	8%	4-13%	11%

CI indicates confidence interval.

\* Housewives, compulsory military service, etc.

([dagger]) Patients with TOF, TGA, or UVH.

([double dagger]) P &lt; .05.



[section] P <.01.

[parallel] P < .001.

TABLE 3. Marital Status of Patients Compared With Expected Levels

All (n = 2896)

	Patients	95% CI	Expected
Single	1079 37% ([dagger])	35-39%	35%
Married	1043 36%	34-38%	37%
Living together	652 23% [section]	21-24%	20%
Divorced	104 4% [section]	3-4%	7%
Widowed	14 0%	0-1%	1%
Unknown	4 0%		

Cyanotic \* (n = 266)

	Patients	95% CI	Expected
Single	132 50%	44-56%	45%
Married	67 25%	20-30%	29%
Living together	64 24%	19-29%	21%
Divorced	3 1% ([double dagger])	0-2%	5%
Widowed	0 0%	0%	

Men (n = 1188)

	Patients	95% CI	Expected
Single	552 46%	44-49%	44%
Married	360 30%	28-33%	31%
Living together	245 21%	18-23%	19%
Divorced	29 2% [section]	2-3%	6%
Widowed	1 0%	0-0%	0%
Unknown	1		

Men (n = 136)

	Patients	95% CI	Expected
Single	79 58%	50-66%	51%
Married	30 22%	15-29%	26%
Living together	27 20%	13-27%	19%
Divorced	0 0% ([dagger])		4%
Widowed	0 0%		0%

Women (n = 1708)

	Patients		95% CI	Expected
Single	527	31%	29-33%	29%
Married	683	40%	38-42%	41%
Living together	407	24% [section]	22-26%	20%
Divorced	75	4% [section]	3-5%	9%
Widowed	13	1%	0-1%	1%
Unknown	3			

Women (n = 130)

	Patients		95% CI	Expected
Single	53	41%	32-49%	38%
Married	37	29%	21-36%	33%
Living together	37	28%	21-36%	22%
Divorced	3	2%	0-5%	6%
Widowed	0	0%		1%

CI indicates confidence interval.

\* Patients with TOF, TGA, or UVH.

([dagger]) P < .05.

([double dagger]) P < .01.

[section] P < .001.

TABLE 4. Number of Patients Having Offspring and Recurrence of CHD Compared With Expected Rates

	n		Having Offspring	
	Patients			
All				
Women	1708	908	53%	([dagger])
Men	1188	441	37%	([dagger])
All	2896	1349	47%	([dagger])
Cyanotic *				
Women	130	45	35%	([dagger])
Men	136	39	29%	([dagger])
All	266	84	32%	([dagger])
			95% CI	Expected
All			51-56%	56%
Women			34-40%	40%

Men	45-48%	49%	
All			
Cyanotic *	26-43%	46%	
Women	21-36%	33%	
Men	26-37%	39%	
All			
	Number of Offspring		
	All	With CHD	
All	1824	50	2.7% * [section]
Women	873	14	1.6%
Men	2697	64	2.4% * [section]
All			
Cyanotic *	79	0	
Women	78	1	1.3%
Men	157	1	0.6%
All			
	95% CI		
All	2.0-3.5%		
Women	0.8-2.4%		
Men	1.8-2.9%		
All			
Cyanotic *			
Women	0.0-3.8%		
Men	0.0-1.9%		
All			

CI indicates confidence interval.

Expected rate of offspring with CHID was considered to be 1%.

\* Patients with TOF, TGA, or UVH.

([dagger]) P < .05.

([double dagger]) P < .01.

[section] P < .001.

Fig. 1. Number and proportion of patients who answered in each diagnostic group.

	all patients	patients who answered
PDA	1066	78%
COA	456	76%
ASD	456	72%
VSD	314	82%

TOF	180	79%
TGA	64	72
PS	123	73%
AS	37	82%
UVH	22	79%
misc	178	73%

#### ACKNOWLEDGMENTS

This study was financially supported by the Finnish Slot Machine Association, Sydanlapset (Parent Organization for Children with Heart Disease), Finnish Foundation for Cardiovascular Research, and Finnish Foundation for **Pediatric** Research.

We thank Hanna Oksanen, PhD, for the statistical review.

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Received for publication Sep 23, 2002; accepted Mar 6, 2003.

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**Record Number:** A111932585

# Pediatric Surgery

**Comment [12]:** I am interested in learning more about pediatric surgery

## Definition

## Purpose

## Demographics

## Description

## Resources

## Definition

**Pediatric surgery** is a specialized field of **surgery** for the treatment of conditions that can be surgically corrected in a baby, child, or adolescent.

## Purpose

The purpose of **pediatric surgery** varies with the procedure. In general, the purpose is to surgically **correct a congenital condition**, disease, traumatic injury, or other disorder in the **pediatric** patient.

**Comment [13]:** most interested in

## Demographics

**Pediatric** surgeons provide treatment for young patients—newborns up through late adolescence.

## Description

**Pediatric surgery** is the surgical branch that uses operative techniques to correct certain **pediatric** conditions (i.e., congenital abnormalities, tumors, chronic diseases, and traumatic injuries). There are different specialties within the field that include:

- **pediatric** general **surgery**



- **pediatric** otolaryngology (ear, nose, and throat)
- **pediatric** ophthalmology (eye)
- **pediatric** urology (urogenital system)
- **pediatric** orthopedic (bone) **surgery**
- **pediatric** neurological (brain and spinal cord) **surgery**
- **pediatric** plastic (reconstructive and cosmetic) **surgery**

The American Academy of **Pediatrics** has established specific guidelines for referral to subspecialists. The **pediatric** patient has special considerations that differentiate him or her, both physically and psychologically, from an adult. A neonate (newborn) poses great challenge in surgical treatment since the tiny structures and immature organ systems may not cope with disease-induced **stress** and the physical demands of a major operative procedure. A newborn infant may still be developing key bodily functions, or may have special requirements. Key areas of concern in the newborn include:

- cardiovascular (heart) system
- thermoregulation (temperature requirements of 73°F [22.8°C]).
- pulmonary (lung) function
- renal (kidney) function
- immature immunity and liver
- special requirements for fluid, electrolyte (necessary elements such as sodium, potassium, and calcium) and nutrition

The **pediatric** surgeon must take into account the special requirements unique to the young surgical patient. The **pediatric** surgeon is trained to treat the entire spectrum of surgical illnesses. The following is an overview (with symptoms) of the more common **pediatric** conditions that require **surgery** typically performed by the **pediatric** surgeon.

### **Alimentary tract obstruction**

Obstruction of the alimentary tract (tubes of digestion extending from the mouth to the anus) is characterized by four cardinal symptoms:

- abdominal distention (an abdomen that becomes large and appears swollen)
- bilious vomiting (due to bile in the stomach)
- maternal polyhydramnios (excess amniotic fluid in the amniotic sac, greater than 2,000 ml) before birth
- failure to pass meconium (dark green or black sticky excretion passed via the newborn's rectum) in the first 24 hours of life

**ESOPHAGEAL ATRESIA AND TRACHEOESOPHAGEAL FISTULA** . This is a congenital

deformity of the esophagus (the tube that passes food from the mouth to the stomach) does not connect to the stomach. Symptoms include severe respiratory distress (the neonate cannot breathe) and excessive salivation. Other clinical signs include **cyanosis** (bluish discoloration of the skin due to oxygen deprivation), **choking**, and coughing.

**PYLORIC ATRESIA AND RELATED CONDITIONS**. Pyloric atresia is a condition that occurs when the pyloric valve, located between the stomach and duodenum, fails to open. Food cannot pass out of the stomach, resulting in **vomiting** clear gastric juice at attempted feedings. Maternal polyhydramnios is present before birth in more than 60% of cases.

Other areas of the colon (duodenum, jejunum, ileum) can be obstructed during development, with symptoms present at birth. Most of these disorders share the four cardinal symptoms of alimentary obstruction.

**INTUSSUSCEPTION**. **Intussusception** accounts for 50% of intestinal obstruction in patients who are three months to one year of age. Eighty percent of cases are observed by the child's second birthday. The cause of intussusception is not known, and it is more common in males who are well nourished and apparently healthy. The symptoms include a sudden onset of **abdominal pain** characterized by episodic screaming and drawing up of the legs. In 60% of patients, vomiting and **blood** in the stool are common findings (either bright red or occult [hidden] blood). Typically, the bowel movements look like currant jelly, consisting of mucus and blood mixed together. Currant jelly stool is the most common clinical observation for patients with intussusception. During **physical examination**, patients will exhibit abdominal distention, and in 65% of cases there is a sausage-shaped mass that can be felt in the upper right portion of the **abdomen** toward the mid-abdomen. Ultrasound studies are a reliable method of diagnosis.

**FAILURE TO PASS MECONIUM**. Failure to pass meconium (meconium ileus) is associated with **cystic fibrosis** (a genetic disorder), colonic obstruction (colonic atresia), meconium plug syndrome, and aganglionic megacolon (also called Hirschsprung's disease, a congenital absence of the nerves that provide gastrointestinal tract mobility).

### **Anorectal anomalies**

There are many different types of anorectal anomalies common to male and female neonates, as well as deformities that are gender-specific since involvement of genitalia can occur. The **surgery** for these cases is complicated, and must be performed by an experienced **pediatric** surgeon. Complications of these procedures could result in permanent problems.

### **Necrotizing enterocolitis (NEC)**

NEC affects 1-2% of patients admitted to a neonatal intensive care unit. It is a life-threatening

illness characterized by abdominal distention, bilious vomiting, lethargy, **fever** , occult (not obvious) or gross (clearly seen) rectal bleeding. Additionally, affected patients may exhibit signs of **hypothermia** (temperature less than 96.5°F or 35.8°C), bradycardia (slow **heart** rate), abdominal mass (felt during palpation), oliguria, **jaundice** , and episodes of breathlessness (apnea). Survival of NEC **surgery** can be expected for 60-70% of patients.

### **Abdominal wall defects**

Omphalocele is a defect that involves protrusion of abdominal contents into an external sac. This disorder occurs in one per 5,000 births. More than 50% of omphalocele patients have serious genetic deformities involving these body systems: cardiovascular (heart), musculoskeletal (muscle and bones), genitourinary (genital and bladder systems), and central nervous (**brain** and **spinal cord** ) . The overall survival rate for infants with omphalocele varies, and depends on defect size, other associated genetic abnormalities, and age of newborn. (Many infants with omphalocele are premature.) Approximately 33% of patients with omphaloceles do not survive.

### **KEY TERMS**

#### **Atresia—**

Thinning or narrowing of a body passageway.

#### **Large intestine—**

The portion of the colon that includes the cecum; ascending, transverse, and descending sigmoid colon; rectum; and anal canal.

#### **Oliguria—**

Decreased urine production.

#### **Pediatric aged patient—**

The **pediatric** aged patient encompasses several periods during development. The first four weeks after birth are called the neonatal period. The first year after birth is called infancy, and childhood is from 13 months until puberty (between the ages of 12 and 15 years in girls and 13 and 16 years in boys).

#### **Polyp—**

A tumor mass, generally benign and capable of surgical removal.

#### **Pylorus—**

The area that controls food passage from the stomach to the first part of the small intestine (duodenum).

#### **Small intestine—**

The part of the intestines that consists of the duodenum, jejunum, and ileum.

**GASTROSCHISIS** . Gastroschisis is a defect in the abdominal wall to the side (lateral) of the umbilicus. It usually occurs to the right of an intact normal umbilical cord. The cause is unknown.

The bowel protrudes to the outside of the abdomen during intrauterine life (while the embryo is developing inside the uterus). The amniotic fluid has an irritating effect on the exposed bowel, and causes **infection** of the bowels. The problem can be detected by ultrasound studies during **pregnancy** . Some **pediatric** surgeons and obstetricians recommend **cesarean section**

(early elective delivery) to spare bowel trauma. The newborn patients typically require **surgery**, **tube feedings** for three to four weeks, and hospitalization for several weeks. The current survival rate for infants with gastroschisis is greater than 90%.

### **Congenital diaphragmatic hernia (CDH)**

CDH can be diagnosed by the fourth month of pregnancy via ultrasound studies. Of the infants with congenital diaphragmatic **hernia** (CDH), 44-66% have other congenital abnormalities as a result of developmental malformations. Anatomically, patients with CDH have a defect in development that allows a communication between the chest and abdomen. Through this defect, the abdominal contents enter the lung cavity and interfere with normal lung development. The incidence is approximately one per 2,200 live births, and males are more commonly affected than females. Usually the infants are full-term, and the defect occurs on the left side in the majority—88%—of patients.

Treatment is extensive, and usually requires three major areas:

- stabilization of patient and preoperative preparation
- operative treatment
- postoperative respiratory, metabolic, circulatory and nutritional supportive measures

Postoperatively, the infant is monitored in the neonatal intensive care setting. The postoperative period is more critical if a lung is severely underdeveloped.

### **Pyloric stenosis (PS)**

Pyloric stenosis is an obstruction in the intestine due to a larger-than-normal size of the muscle fibers of the pylorus (lower stomach opening). Pyloric stenosis is a common hereditary condition that affects males more than females, and occurs in one per 750 births. The typical symptoms include a progressive, often projectile, vomiting after attempted feedings. The gastric vomitus (bloody in 80% of patients) usually begins during the second and third week of life, and increases in force and frequency. Typically, the infant fails to gain weight, and the number of bowel movements and rate of urination decreases.

Physical examination is usually helpful in establishing a diagnosis. Palpation of the enlarged muscle fibers can be felt as an olive-shaped mass located along the midline approximately one-third to one-half of the distance from the umbilicus to the xiphoid (end of the breastbone), when the stomach is empty. Careful abdominal examination and palpation can usually identify the pyloric mass in 85% of cases.

### **Gastroesophageal reflux**

Gastroesophageal reflux disease (GERD) is a common disorder in infancy, and usually disappears by the baby's first birthday. The largest group of patients with clinically significant GERD are those who have neurologic impairment. Symptoms often include vomiting, repeated lung infections (from aspirating gastric contents during regurgitation of foodstuffs), and delayed gastric emptying. The success rate with infants who have procedures necessary to correct GERD is over 90%.

### **Meckel's diverticulum**

Meckel's diverticulum occurs in approximately 2% of the U.S. population. The diverticulum is an outgrowth of intestine that is located in a portion of the intestines called the ileum. Symptoms of obstruction are more often observed in infants, and bleeding is more common in patients after age four.

### **Intestinal polyps**

Juvenile polyps are usually present between the ages of four and 14 years, and tend to be inflammatory. The most common symptom of intestinal polyps is rectal bleeding, which is commonly due to a solitary polyp (80% of cases). Diagnosis can be done by proctosigmoidoscopy, which allows visualization of 85% of polyps.

### **Acute appendicitis**

Acute **appendicitis** is a relatively common surgical emergency that is misdiagnosed in 28% of patients due to a broad spectrum of symptoms that can confuse the clinician. The classic clinical symptom of acute appendicitis is the onset of **pain** in the middle region of the abdomen that is followed by anorexia (loss of appetite), nausea, and vomiting. The pain is persistent and radiates to the right lower abdomen, becoming more intense and localized. The physical and abdominal examinations must be carefully and accurately performed. Patients with acute appendicitis usually have an increased white blood cell (cells that fight infection) count.

Once the diagnosis is established, the child is prepared for **surgery**. Preoperative **antibiotics** are started at least one half-hour before the operation. If the appendix is perforated (ruptured), complications can occur as a result of kidney (renal) failure, seizures due to fever, and gram-negative **sepsis** (an infection that enters the bloodstream and interferes with life-saving chemical reactions). Patients who are very young, or those who were misdiagnosed and incurred long delays in treatment, are susceptible to death.

### **Inflammatory bowel diseases**

Some cases (approximately 25%) of inflammatory bowel disease are found in persons younger than 20 years of age. Two types can occur, **Crohn's disease** and ulcerative **colitis**.

The diagnosis of inflammatory bowel disease is usually based on presenting clinical symptoms, laboratory analysis results, endoscopic appearance, and radiologic findings. Approximately 50-60% of patients have bloody **diarrhea**, severe cramping, abdominal pain, and urgency.

**CROHN'S DISEASE**. The symptoms of Crohn's disease includes cramping abdominal pain, diarrhea, and strictures (constriction) resulting from bowel obstruction. Removal of diseased portions in children with Crohn's disease may be temporarily beneficial, but recurrence after surgical removal occurs in about 50% of cases within four years. Chronic symptoms may remain into adult life, making long-term follow-up essential.

**ULCERATIVE COLITIS**. Ulcerative colitis is limited to the colon. A surgical procedure known as colectomy is curative, and indicated for intractable disease (64% of patients). Colectomy is the removal of the entire colon, or the inflamed part of it.

### **Biliary tract disorders**

A variety of biliary tract conditions may be present at birth, some requiring surgical correction.

**NEONATAL JAUNDICE**. **Neonatal jaundice** is common, and results from an immature system not capable of some basic biochemical reactions. Food intake can help speed these reactions, which usually resolve the condition within seven to 10 days. Jaundice that persists for over two weeks is abnormal, and could be caused by over 30 possible disorders.

**BILIARY ATRESIA**. Biliary atresia is a disease that causes inflammation of the ducts within the biliary system, resulting in fibrosis of these ducts. The incidence of biliary atresia is one per 15,000 live births, and is more common in females. Time is critical, and most patients must have **surgery** by two months of life. Approximately 25-30% of patients who receive early operative intervention have long-term successful outcomes. Some patients may require **liver** transplantation, and 85-90% of these patients survive.

**CHOLELITHIASIS**. **Gallbladder** obstruction in infants and young children is usually caused by pigmented (colored) stones resulting from blood disorders. Removal of the gallbladder (laparoscopic cholecystectomy) is the treatment of choice.

### **Trauma**

Accidents are the leading cause of death in children between the ages of one and 15 years, and accounts for 50% of all deaths in the **pediatric** age group. More than half of these deaths are due to motor vehicle accidents, followed by **falls**, bicycle injuries, drowning, **burns**, **child abuse**, and birth trauma. Head trauma is the single most common organ associated with

traumatic death. Within recent years, the number of fatalities related to the use of firearms and violence has increased.

### **WHO PERFORMS THE PROCEDURE AND WHERE IS IT PERFORMED?**

**Pediatric surgery** is performed by a **pediatric** surgeon who has had five years of general **surgery** training, along with further specialized instruction and experience, and is certified in **pediatric** general **surgery** or in a specific **pediatric** specialty.

More than 20 million children each year sustain injuries requiring treatment. These injuries account for 100,000 cases of permanent **pediatric** disability. Response to trauma in **pediatric** patients is significantly different from older patients. **Pediatric** patients require special attention concerning temperature regulation, blood volume, metabolic rate and requirements, and airway maintenance. Other special **pediatric** considerations include response to stress, communication difficulties, psychological trauma, a different **pediatric** trauma score system, smaller airway diameter, and increased risk of aspirating gastric contents (which could cause **pneumonia**). **Pediatric** trauma patients should have access to appropriate pre-hospital transportation, and must receive medical attention in a **pediatric** trauma center capable of providing the complex level of care necessary for serious **pediatric** trauma situations.

### **Neck masses**

Neck masses during infancy and childhood may be caused by tumors or infections, or they may be congenital. Lymphadenitis is an infection of a lymph node that becomes enlarged and tender. Most cases are resolved by treating the primary source of infection (i.e., middle ear infection and **tonsillitis**). Some inflamed nodes may require an incision and drainage of infection.

### **Hernias**

**INGUINAL HERNIA AND HYDROCELE**. Inguinal (groin) hernia is the most frequent disorder requiring **surgery** in the **pediatric** age group. Clinically, a rightsided inguinal hernia is more common in males (60% of cases), and there is a familial tendency. The incidence is higher in full-term infants (3.5-5%). Full-term infants and older children (without underlying diseases) can receive surgical repair in an outpatient setting. An inguinal hernia may result in herniation of the scrotum, and a communicating hydrocele (hernia with a small connection to the peritoneal cavity).

**UMBILICAL HERNIA**. Umbilical hernia is a defect of the umbilical ring, and is more common in females and African American children. Spontaneous involution occurs in 80% of cases. Larger defects may be observed for several years without complications, and their spontaneous

resolution is possible. If the umbilical hernia persists, patients may develop feeding intolerance, pain, and local skin breakdown.

### Undescended testes

Undescended testes are observed in 1-2% of full-term males. Approximately 30% of preterm males may have an undescended testis. Undescended testis in [premature infants](#) may descend by the first year of life, and observation is often the treatment during that time.

### Tumors

Wilm's tumor (nephroblastoma) is a tumor in the [kidneys](#) that forms during embryonic development. The tumor is due to a genetic abnormality; and approximately 80% of children are diagnosed between one and five years of age. In about 75-95% of cases, the patient has an abdominal mass that is detected by a parent during bathing. Blood in the urine (hematuria) occurs in 10-15% of cases, and high [blood pressure \(hypertension\)](#) is present in 20-25% of cases. Hypertension is the result of the tumor compressing the kidney in a specific area, causing it to release a chemical called renin, which elevates blood pressure. During physical examination, the Wilm's tumor is a smooth, round, hard, nontender flank mass. The treatment of Wilm's tumor depends on its stage, and may include [surgery](#), [chemotherapy](#), or [radiotherapy](#).

### QUESTIONS TO ASK THE DOCTOR

- What are the risks of [surgery](#)?
- What is the benefit of the [surgery](#)?
- What type of anesthesia will be used?
- How many surgeries of this type has the surgeon performed?
- Is there an alternative to [surgery](#)?
- Is a full recovery expected, and if not, what deficits will the child have?
- What should the parents and child do to prepare for [surgery](#)?
- What care is needed following the [surgery](#)?
- When will the child be able to return to normal activity?

### Resources

#### BOOK

Townsend, Courtney. *Sabiston Textbook of [Surgery](#)*, 16th ed. St. Louis: W. B. Saunders Company, 2001.



## PERIODICALS

Coran, A. "American Academy of Pediatrics: Guidelines for Referral to **Pediatric** Surgical Specialists." *Pediatrics* 110, no. 1 (July 2002).

Okada P. J., B. Hicks. "**Pediatric** Surgical Emergencies: Neonatal Surgical Emergencies." *Clinical**Pediatric** Emergency Medicine* 3, no.1 (March 2002:).

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**Record Number:** CX2760400814