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Pectus excavatum in a 112-year autopsy series: anatomic findings and the effect on survival

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Abstract

Background/Purpose: The purpose of this study was to determine the frequency of pectus excavatum and associated conditions in a large autopsy series. It also sought to determine whether there were different survival patterns for pectus excavatum patients than for patients without pectus excavatum.

Methods: A computer-assisted search of autopsy files maintained by Johns Hopkins University was conducted, dating from 1889 to 2001. Each patient's Autopsy Pathology Information System report was reviewed for diagnosis and comorbid conditions. To determine whether there were differences in survival patterns, we tested whether pectus excavatum patients survived longer than controls, using a standard epidemiological method. Each patient in the autopsy series was compared with the 2 patients entered in the autopsy database chronologically immediately before and the 2 patients immediately after the case. A Kaplan-Meier survival analysis was conducted.

Results: Pectus excavatum was identified at autopsy in 62 of 50,496 cases. Of these 62 patients, 17 were 65 years or older and appeared to have died of causes unrelated to pectus excavatum, the oldest being 91 years. Twenty-one were between the ages of 14 and 65 years and were found to have coexisting conditions or syndromes. Six were between the ages of 1 and 4 years. One of the 6 died in 1947 because of complications from pectus repair. No autopsied patient with pectus excavatum died between the ages of 5 and 14 years. Eighteen were infants younger than 1 year, and all 18 died because of conditions unrelated to pectus excavatum. There were no reported cases of pectus excavatum before 1947, and the severity of deformity could not be determined from the autopsy data. Survival analysis indicated that pectus excavatum patients had a different survival than the controls. Pectus excavatum patients tended to die earlier ($P = .0001$). However, pectus excavatum patients who survived past the age of 56 years tended to survive longer than their matched controls ($P = .0001$).

Conclusion: Although there were no histological abnormalities noted in the cartilage of the pectus excavatum patient's conditions, pectus excavatum was associated with several connective tissue abnormalities. Analysis is consistent with the theory that this condition can impact survival.

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Pectus excavatum is a commonly occurring chest wall deformity that often coexists with other disorders, such as connective tissue disorders. The incidence of pectus excavatum and its occurrence with other disorders, however, has not been well established. Thus, one of the reasons for our study was to examine the frequency of pectus excavatum and its associated conditions.

To date, there has been little research into the impact of pectus excavatum on a patient's survival [1]. Prior research has focused on surgical correction of pectus and the associated outcomes, including the resultant improvement in cardiac and/or respiratory functioning. In our analysis, we tested the hypothesis that individuals with pectus excavatum tend to die at an earlier age than do individuals without pectus excavatum.

1. Methods

This study was approved by the Johns Hopkins institutional review board. To determine the frequency of occurrence of pectus excavatum in the patient population, the autopsy files of the Johns Hopkins Hospital were searched for the diagnosis of pectus excavatum and any related terms such as recurvatum and funnel chest. The autopsy files on all cases identified were reviewed for comorbid conditions.

We were also interested in examining whether pectus excavatum had an effect on the survival rate of pectus excavatum patients. Patients identified with pectus excavatum were matched with 4 controls consisting of the 2 race- and sex-matched patients autopsied before and the 2 after the pectus excavatum patients. The only data available on the matched controls were age at death, date of death, and pectus status. Survival analyses included only those patients who were age-eligible for surgery. Because we wanted to assess whether surgical repair could potentially affect survival, and because surgery is not presently performed on patients younger than 3 years, this age group was not included in the analysis.

To determine whether there were differences in survival patterns, we plotted Kaplan-Meier survival curves for the pectus excavatum cases and the controls as a group and tested the significance of differences using Wilcoxon weight log-rank tests to account for lack of proportionality in the hazard curves. We assessed interaction by examining the plots to see if they crossed and stratified at the point of interaction for statistical tests of significant differences in survival pattern. All analyses were conducted using Proc Lifetest in SAS 8.0 (SAS, Cary, NC).

2. Descriptive findings

Of the 50,496 autopsy files reviewed, pectus excavatum was diagnosed in 62 cases. The ages of the autopsy patients with pectus varied widely, with the oldest being 91 years. Pectus excavatum was confirmed in patients from infancy to age 4 and from age 14 or older. None of the autopsied patients with pectus excavatum died between ages 5 and 14 years. Eighteen patients were younger than 1 year at death, 6 patients were between the ages of 1 and 4 years, 21 patients were between the ages of 14 and 65 years, and 17 patients were 65 years and older. Of the 62 patients, 48 (77%) were male patients.

The severity of the pectus deformity could not be determined from the autopsy data. Computed tomography (CT) scan data were not sought because the number of patients autopsied after CT scanning became available was too small to allow for statistical inferences.

Of the 21 patients who died between the ages of 14 and 65 years, 2 had Marfan syndrome, and there were singular cases of Ehlers-Danlos syndrome, Noonan syndrome, Duchenne muscular dystrophy, and Rett syndrome. One patient had been diagnosed with scleroderma, and another patient had questionable scleroderma. One patient had developed progressive scoliosis despite undergoing pectus repair at age 2 years.

Of the patients in the age group 1 to 4 years, 1 died in 1947 of complications related to pectus repair (wound infection and right empyema).

Of the 18 patients younger than 1 year, 2 had Werdnig-Hoffman disease, 1 had infantile muscular dystrophy, and 1 had Zellweger cerebrohepatorenal syndrome. One of the infants had spina bifida and underwent pectus repair but died in 1955. Nine of the infants had pneumonia at autopsy, and an additional 3 had hyaline membrane disease.

The earliest occurrence of pectus excavatum in the autopsies was in 1947. There were no noted cases in the first 20,400 autopsies ranging from 1889 to 1947. The median age at death for the pectus excavatum patients was 33 years (interquartile range 0-65 years) and for the controls was 44 years (interquartile range 3.5-65 years). Fig. 1 illustrates

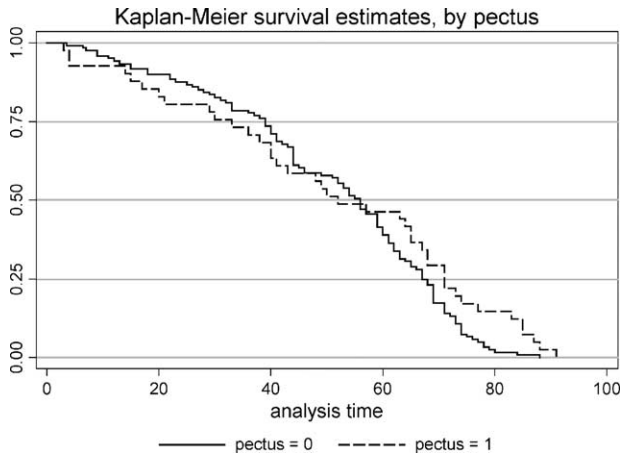

Fig. 1 Proportion of patients surviving to given age among individuals who survived to age 3 years according to pectus excavatum status.