

higher rate of urological anomalies within the spectrum and a higher rate of heart, abdomen, skeleton, and joint anomalies outside the spectrum. These might be either important in terms of predictable comorbidity for preparing the primary reconstruction in the newborn or infant period or later in the long-term follow-up. Their clinical relevance suggests considering them before initial operative treatment. As any associated anomalies were present in 48 up to 59% of affected EEC individuals possibly associated anomalies should be evaluated during routine check-up in all EEC patients.

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EDITORIAL COMMENT



The authors present a large Multicenter German Study looking at the association between the exstrophy epispadias complex (EEC) and other congenital anomalies. The database was created from patient's entering the CURE-net system, which started in 2009. In the prospective group, the authors have identified 73 patients born between the years of 2009 and 2016. They also identified a larger group of 162 patients which they label as a cross-sectional group who were recruited from EEC born between 1948 and 2008. By far the most valuable of the groups is the prospective group which includes the 73 patients starting in 2009.

The authors' series does point out several intriguing and interesting associations some of which are different from previously published reviews of anomalies associated with the EEC. The first of these would be that exstrophy variants were found in 21% of the current review, which is significantly higher than previously reported. The authors found that associated anomalies outside the EEC were discovered, in 59% of their prospective group including renal anomalies in 18% and genital anomalies other than the epispadias component in 18%. They also pointed out that females in their study group were found to have Mullerian duct abnormalities to a far greater extent than had previously been reported with 65.7% experiencing Mullerian duct abnormalities such as uterine or vaginal duplication obstruction or absence.

Although the authors' data is compelling, it may not give the full picture due to a lack of mandatory reporting congenital anomalies through a centralized birth registry in German. The number of pregnancies in which the EEC was discovered and terminated before birth are not documented and no further valid data are available such as other anomalies which might have been present. Although, much of the data is intriguing and thought provoking, I am not entirely sure that it in any way changes our approach to the care of the infant born with EEC. Most of these patients are completely investigated, thus few, if any of the anomalies reported would be inadvertently overlooked.

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AUTHOR REPLY



We greatly appreciate the Editorial Comment on our article “Association Between Exstrophy-Epispadias Complex And Congenital Anomalies: A German Multicenter Study.” Here the question has been raised whether the observations of our study

might change the approach to the care of newborns with exstrophy-epispadias complex (EEC).

In our study, we compared 2 EEC cohorts. The prospective cohort comprised patients approximately 1 year old at the time of data acquisition and a cross-sectional cohort of older EEC patients. Patients in the prospective cohort were born between 2009 and 2016. Patients in the cross-sectional cohort were born between 1948 and 2008. We observed more exstrophy variants and co-occurring congenital heart defects in our prospective cohort compared to our cross-sectional cohort ($P < .0001$ and $P = .003$, respectively). Moreover, we observed more co-occurring congenital anomalies in our prospective cohort compared to our cross-sectional cohort, although this difference was not significant ($P = .16$).

While exstrophy variants constitute very rare orchids within the EEC spectrum, we believe that only the exact preoperative assessment of these exstrophy variants guarantees the best operative care. Furthermore, the correct assessment of the cardiovascular status prior to major reconstructive surgery will reduce the congenital heart defects associated perioperative risks. Hence, stage of the art treatment should encounter the exact assessment of the patients' phenotype, including all co-occurring congenital anomalies.

In this respect, we would like to quote the German poet and philosopher Johann Wolfgang von Goethe [1749-1832] who said "We only see what we know."

We deliberately advocate that if prenatal diagnosis raises the suspicion of an EEC phenotype, or when a newborn presents with anomalies of the EEC spectrum, the respective parents or newborns should be forwarded to a specialized pediatric urology department for further consultation and ultimate operative treatment.

Prior to an immediate postnatal transfer, we urge the primary physician to get in touch with a specialized pediatric urology department to receive consultation for the initial assessment. We urge these physicians to seek advice on the opportunities for initial conservative treatment regimens including the coverage of the bladder plate with a sterile gauze, allowing the newborn and the mother to bond instead of separating an otherwise healthy newborn from his mother.

Up to now, we are certain that even if Germany had a nationwide centralized birth registry, the primary physician who assesses the fetus prenatally or the newborn at birth could only then register the case with an anomaly of the EEC spectrum "if he knows what he sees."

We believe, that only the thorough assessment of a newborn with a complex urogenital anomaly in a specialized pediatric urology department will guarantee the best care, a matter of course, far from the current practice.

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